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THE ELECTROMYOGRAM IN MYOPATHY: ANALYSIS WITH THE AUDIO-FREQUENCY SPECTROMETER

BY

JOHN N. WALTON

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Despite the fact that in most established cases of muscular dystrophy the clinical diagnosis is self-evident, occasional cases arise in which it is difficult to decide whether muscular wasting is of neuropathic or myopathic origin. Careful work by Kugelberg (1949) and others has shown that electromyography may be of considerable value in making this fundamental distinction, but in many centres the techniques for diagnosis are not yet firmly established. In this paper the electromyographic findings in 100 cases of myopathy are reviewed, and a method of frequency analysis is described which appears to enhance the value of the electromyogram in diagnosis. The results obtained by applying this analytical technique in the examination of 26 normal subjects and 48 patients with other neurological disorders are compared with those noted in the cases of myopathy. Before discussing the methods and results of the investigation, however, it is essential to discuss briefly the physiological principles involved and the findings of other authors.

The unit of muscle structure is the muscle fibre; under normal conditions muscle fibres never contract singly, but single fibre or fibrillation potentials can be recorded from resting muscle after denervation in the form of monophasic or diphasic spikes (Fig. 1) of about 1 millisecond duration (Weddell, Feinstein, and Pattle, 1944). Normal volitional activity of voluntary muscle is made up by the asynchronous contraction of individual motor units; each unit consists of the bundles of muscle fibres supplied by one anterior horn cell and its motor neuron. Electrically, the activity of the fibres of the unit summates smoothly to give a monophasic, diphasic, or triphasic wave, the motor unit action potential (Fig. 2), which varies between 4 and 10 milliseconds in duration (Petersén and Kugelberg, 1949). Hence it will be seen that in sustained contraction of voluntary muscle the dominant frequency of the electrical discharge, depending on the duration of the individual waves from which the complex wave form of the interference pattern is synthesized, will be of the order of 100 to 250 cycles per second. In disease of the anterior horn cell and the proximal part of the lower motor neuron it is clear that whole motor units, supplied by the diseased neuron, will perish, and for this reason the interference pattern will be reduced; eventually after the death of many neurons a needle electrode may record the activity from isolated surviving units, the "single discharge" of Buchthal and Clemmesen (1941). Under these circumstances, since the remaining motor units are of normal duration, the dominant frequency of the wave form will be unchanged.

In primary disease of the muscle, on the other hand (e.g. muscular dystrophy), no such anatomical pattern of degeneration obtains and individual muscle fibres die sporadically. No longer, therefore, is it possible for the activity of the fibres of a motor unit to summate smoothly, as some components of the unit will be missing, and for this reason many polyphasic potentials are seen (Fig. 3). If degeneration has proceeded further it may be that one or two fibres are all that remain of a motor unit and fibrillary potentials may be obtained on volition. Hence it will be expected that the dominant frequency of the electrical discharge from contracting dystrophic muscle will approach more nearly to 1,000 cycles per second. Kugelberg (1949) examined 16 cases of muscular dystrophy and carried out careful measurements of the motor unit potentials recorded. These findings were compared with those in 142 cases of neuropathy and it was confirmed that in the cases of myopathy a greatly increased proportion of polyphasic potentials and attenuated or short duration potentials was produced on contraction of the biceps brachii.

It should be remarked at this juncture that a number of polyphasic potentials are to be found on recording from normal muscle (Denslow and Hassett, 1943; Weddell and others, 1944); the proportion is about 2 to 4% in the biceps

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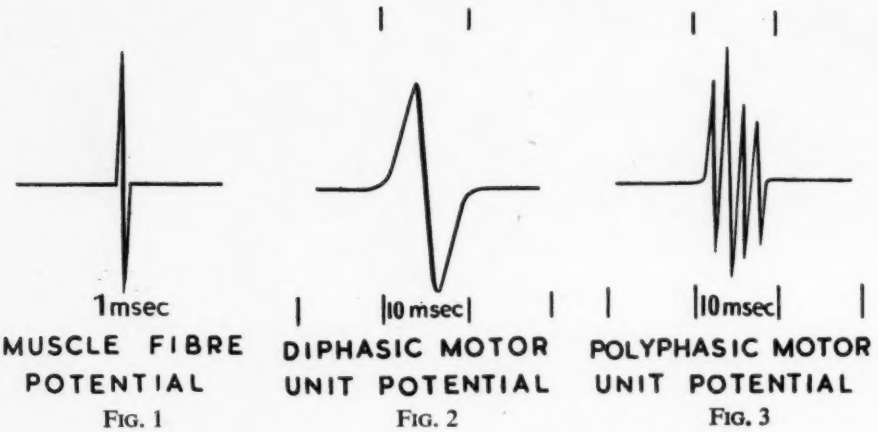

FIG. 1 FIG. 2 FIG. 3

FIG. 1.—Diagrammatic representation of a muscle fibre potential (amplitude about 100 microvolts).

FIG. 2.—Diagrammatic representation of a biphasic motor unit potential (amplitude about 1–2 millivolts).

FIG. 3.—Diagrammatic representation of a polyphasic, partially disintegrated, motor unit potential as seen in myopathy (amplitude about 500 microvolts).

muscle and considerably higher in the facial musculature (Petersén and Kugelberg, 1949). Similar disintegrated motor unit potentials also occur in large numbers during regeneration after a peripheral nerve injury, owing to the irregular reinnervation of fibre groups; they are also occasionally seen in cases of distal neuropathy where the degenerative process affects the nerve fibres distal to the point of branching of the motor neuron, when the scattered muscle fibres of the motor unit may degenerate at different rates (Richardson, 1951a, 1952). Fortunately the latter eventuality is uncommon, occurring only in occasional cases of polyneuritis, and the proportion of polyphasic potentials is not usually as great as that noted in cases of myopathy.

Other diagnostic features of the electromyogram in muscular dystrophy have been discussed by Kugelberg (1949) who pointed out that spontaneous fibrillation is absent in the myopathies but is often present in neuropathy; furthermore a sustained interference pattern is often readily obtainable from grossly wasted muscle in myopathy, whereas in the neuropathies the pattern, and in particular, the number of spikes, is usually reduced in proportion to the diminution of muscular power.

From the above information there has emerged a characteristic electromyogram of myopathy. The muscle is usually silent at rest, but on volition, even in extremely weak and wasted muscles, a highly complex polyphasic interference pattern will be produced, and the noise in the loudspeaker is high-pitched, like hail on a tin roof. In neuropathy, on the other hand, there is often spontaneous fibrillation with the muscle at rest, and on volition the interference pattern is interrupted or reduced. The loudspeaker gives an intermittent, low-pitched rumble, save in occasional cases where the neuropathic degeneration is predominantly distal, when a certain number of polyphasic potentials, with their high-pitched, crackling note, may be added.

However, although the characteristic findings described above are almost invariably noted in established cases where the clinical diagnosis has been made with ease, it is in the cases which give rise to diagnostic difficulty clinically that the electromyogram may also give equivocal results. The presence of a small proportion of polyphasic potentials may obscure the underlying potentials of normal duration; on the other hand the observer may not consider that such high frequency activity is present in sufficient quantity for a firm diagnosis. Visual analysis of the ephemeral trace on a cathode ray screen is notoriously unreliable; admittedly it is possible to submit the photographed record to detailed visual or mathematical analysis, but this procedure is time-consuming, and certainly any complex wave-form may contain components which are invisible to the eye (Dawson and Walter, 1944). Furthermore, sampling and measurement of individual motor units is also laborious and it is impossible to be certain that the recorded data are representative of the whole muscle.

For these reasons it was decided to seek a convenient method of instantaneous frequency analysis of the electromyogram. Recently Richardson (1951b) has described a simple technique; his analyser is based on electronic filters and integrating voltage meters which filter off the frequency components above 400 cycles per second and measure them. He has suggested that an increase in the measurement of these high frequencies compared with those below 400 cycles indicates a myopathic lesion, but his method has not yet been applied to a large series of cases. In 1946 Agate and Druett reported the results of an investigation in which an audio-frequency spectrometer was utilized to study vibrations transmitted to the hands; since the frequency spectrum covered by this instrument

seemed satisfactory it was decided to employ it as an analyser of the electromyogram.

Material

One hundred cases of myopathy have been examined; a clinical study of these cases is to be reported in detail elsewhere. The cases are listed according to the clinical type of the disease in Table I; the diagnoses of other cases examined by this method, for comparison, are also given.

TABLE I
ANALYSIS OF CASES EXAMINED

	Diagnosis	No. of Cases
Cases of Myopathy	Pseudohypertrophic muscular dystrophy	43
	Facio-scapulo-humeral muscular dystrophy (Landouzy-Déjerine)	12
	Late juvenile muscular dystrophy (Erb's scapulo-humeral type)	19
	Dystrophia myotonica	15
	Muscular dystrophy of late onset (Nevin)	3
	Distal myopathy of Gowers	1
	Ocular myopathy	1
	Atypical myopathy	6
	Total	100
Other Cases	Normal controls	26
	Motor neuron disease	13
	Polyneuritis	4
	Neuralgic amyotrophy	2
	Dermatomyositis	2
	Myotonia congenita (with no evidence of muscular wasting)	6
	Amyotonia congenita (Oppenheim)	1
	Peripheral nerve injury	8
	Facial paralysis	9
	Cervical spondylosis	3
	Total	74

Methods

All records were taken using standard concentric needle electrodes and the "stanco" double channel electromyograph. For the investigation only one channel of the apparatus was required and the output of this channel was fed into the audio-frequency spectrometer. A photograph of the apparatus is given in Fig. 4.

For technical details of the spectrometer the reader is referred to the paper by Agate and Druett (1946). Briefly, it consists of 27 resonant circuits tuned to frequencies distributed logarithmically throughout the 40–16,000 c.p.s. range, each having an associated storage condenser. These are scanned sequentially every 1/24 of a second by a rotary switch and their accumulated voltages are fed first into a modulator and thence on to the Y-plates of a cathode ray oscilloscope. Incremental X-deflections provided by the switch itself combine to produce on the screen a frequency/volt × second histogram. At rest the screen shows a row of dots each corresponding to a particular frequency (Fig. 5); when any wave form is fed into the instrument vertical lines appear on the dot of the responding frequencies, the amplitude of each line being proportional to the amount of the frequency concerned. Care must be taken in adjustment of the gain control to prevent overloading,

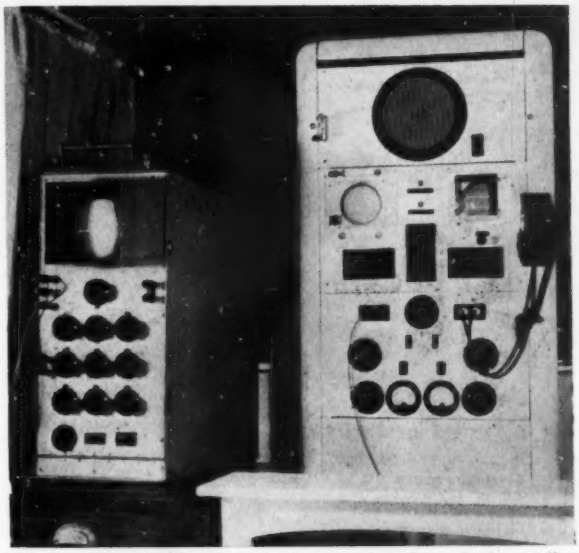

FIG. 4.—The double channel electromyograph and the audio-frequency spectrometer (on the left).

since when this occurs excessive responses may be produced at harmonics of the responding frequencies, and this may encourage erroneous interpretation. With experience, this error is easily avoided.

Except in young children, where needling of muscle was ill-tolerated, at least three separate muscles were examined in every case of myopathy, each with a single insertion. Two or more muscles were examined similarly in the controls and other patients. Extensive probing was then carried out and the electromyogram, with a simultaneous analysis, was obtained first at rest and then on volition, with the needle point in at least five separate positions within the muscle. Direct visual and auditory analysis was relied upon for interpretation of the electromyogram, but the spectrometer histogram was invariably photographed, either with a 16 mm. "ciné kodak" camera supplied with the apparatus, or occasionally by making repeated single exposures on 35 mm. film with a "leica". For statistical reasons, extensive sampling of individual muscles, using several insertions, with the needle point at standard depths within the muscle, was envisaged, but the practical difficulties were so great and so many variable factors were involved that this plan was discarded. Furthermore it soon became clear from the unequivocal results obtained that the added discomfort occasioned to patients by repeated and extensive explorations was both unnecessary and unjustifiable. As it was also clear that the analysis was unaffected (except in the amplitude of its components) by increasing activation, no account was taken of the power of contraction of individual muscles, but as a rule the record was taken during maximum voluntary effort against resistance.

Results

Normal Controls.—Before any patients with myopathy or other neurological disorders were

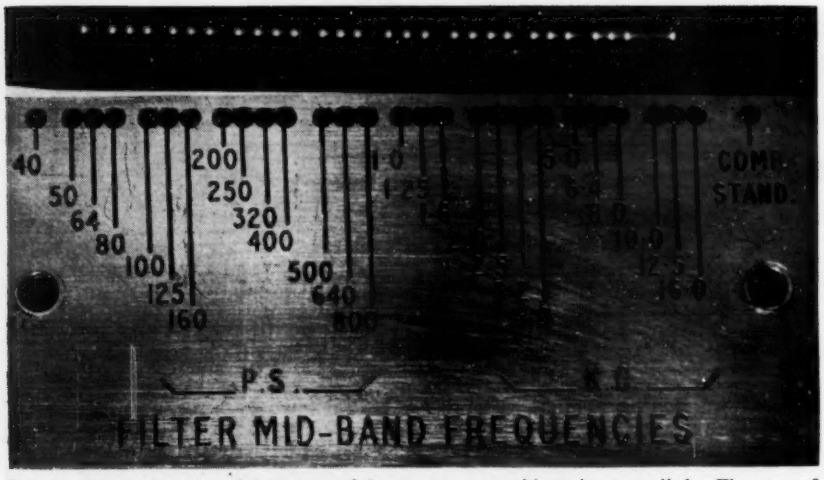

FIG. 5.—The appearance of the screen of the spectrometer with no input applied. The row of dots described in the text are seen, and a key is superimposed indicating the frequencies corresponding to each dot.

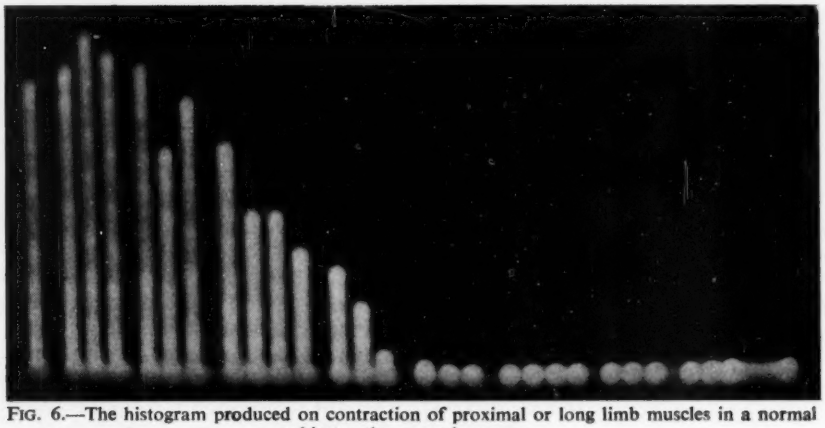

FIG. 6.—The histogram produced on contraction of proximal or long limb muscles in a normal subject: the normal response.

examined a total of 36 examinations were made on 26 control subjects, 16 males and 10 females. Although most of the subjects were junior medical staff or students and hence in the third or fourth decades the results obtained in the few children and elderly people examined did not suggest that the frequency analysis varied significantly with age. Three separate examinations in different subjects were made of each of the following muscles: orbicularis oris, masseter, deltoid, pectoralis major, biceps brachii, brachioradialis, flexor pollicis brevis, adductor pollicis, gluteus maximus, quadriceps femoris, gastrocnemius, and tibialis anterior. In all the large limb muscles the spectrometer histogram obtained when the muscle contracted was quite uniform, showing a peak frequency response at 100–200 cycles per second, and tailing off to zero at about 800 cycles. Fig. 6 is an example of the pattern obtained from the deltoid muscle of a normal subject; this standard response will hereafter be referred to as the normal pattern. The pattern did not vary when the needle was moved into many different parts of the muscle, nor did it depend on the degree of activation.

A significantly different histogram was produced on examination of the masseter and orbicularis oris; the dominant frequency in these situations was at 200–250 c.p.s. and the spectrum was broadened by the accession of higher frequencies, with some response even at 1,500 c.p.s. Fig. 7 is an example of this type of histogram, which will be referred to as the broad spectrum type of response. A spectrum of this type was also obtained frequently from the flexor pollicis brevis and has since been observed in recordings from other small hand muscles in normal subjects. These observations can be correlated with the fact already mentioned that a high proportion of polyphasic potentials can be obtained from the normal facial musculature; it appears that the same findings apply to the small muscles of the hand.

Myopathy.—It will be convenient to consider first the results of frequency analysis, and then other electromyographic findings in these cases.

Frequency Analysis.—On examination of the first case it was immediately evident that on recording from muscles severely affected by the dystrophic process a spectrometer histogram could be obtained which differed significantly from any of the findings in normal subjects. In this pattern there was a relative absence of the lower frequencies and there was a complete shift to the right of the dominant frequency. An example is given in Fig. 8, which shows the histogram obtained from the contracting pectoralis major of a patient with facio-scapulo-humeral muscular dystrophy. It will be seen that the dominant frequency response is at 640 c.p.s., and the response at 100–250 c.p.s. is compara-

tively scanty. This pattern will be referred to as the shift type of histogram. It has been considered that a significant shift occurred when the dominant frequency exceeded 400 c.p.s. Such a pattern was never seen in any situation, even after extensive probing, in the normal subjects examined. Detailed results are given in Table II.

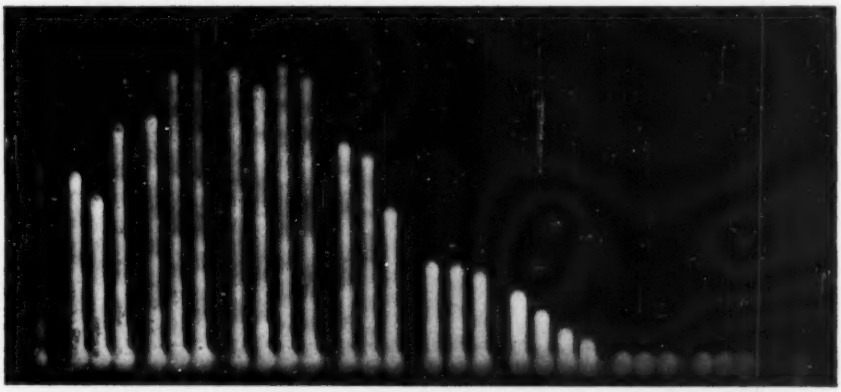

FIG. 7.—The histogram produced on contraction of facial muscles or small hand muscles in a normal subject: the broad spectrum type of response. A similar pattern is obtained on examination of relatively unaffected muscles in patients with myopathy.

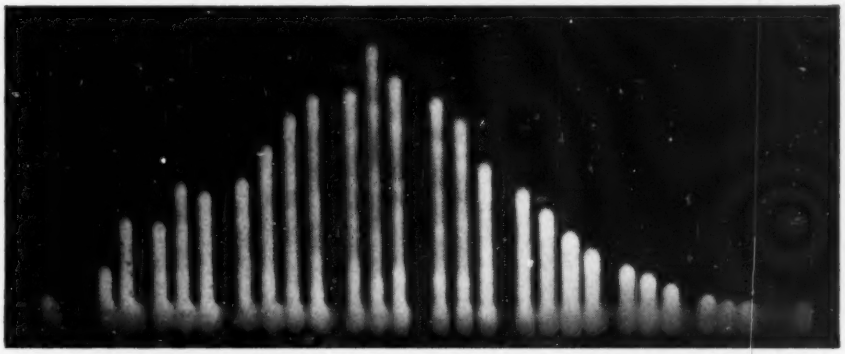

FIG. 8.—The histogram produced on contraction of the pectoralis major in a patient with facio-scapulo-humeral muscular dystrophy, showing a shift to the right of the dominant frequency: the shift type of response.

In 98 of the 100 cases examined it was possible to obtain a shift type of analysis from at least one of the muscles examined. In some cases, particularly those in which the disease process was advanced, such a pattern was seen on volition whenever the needle was randomly inserted into the muscle. On other occasions some probing was necessary before a shift occurred, the pattern from the remainder of the muscle being occasionally normal (in early cases), but more often giving a broad spectrum response. It was noted that in such cases a shift was most commonly obtained on recording from the more superficial portions of the muscle, suggesting that degeneration was more advanced in this situation. The two patients in whom no shift was obtained were young children in whom the examination was of necessity brief and incomplete; in both cases the analysis was abnormal, a broad spectrum response being obtained from long limb muscles, but it is probable that a shift would have occurred with more extensive probing.

Consideration of the results listed in Table II will show that as would be expected a shift was most readily obtained from the muscles most affected by the disease process. Thus in dystrophia myotonica, where the myopathic process is predominantly distal in distribution, the characteristic findings were obtained from forearm muscles and those below the knee. In the other forms of muscular dystrophy, where proximal muscles are chiefly involved, the most definite results were obtained from examination of the pectoralis major and biceps brachii in the upper limbs and quadriceps and tibialis anterior in the lower. A parallel clinical survey has shown that the spinati, deltoid, and gastrocnemius may remain relatively powerful until a late stage of the disease in these cases, and it will be seen that the pattern of analysis was generally less abnormal when these muscles were examined. Hence it is suggested that for the examination to be of most value the pectoralis major, biceps brachii, quadriceps, and tibialis anterior should be examined in cases of muscular dystrophy of the proximal type, while in dystrophia myotonica and distal myopathy attention should be turned to the long flexors and extensors of the fingers and tibialis anterior.

Spontaneous Activity.—Kugelberg (1949) has stressed that the absence of fibrillation is in favour of the diagnosis of myopathy; spontaneous fibrillary potentials were not observed in any case of the present series. On several occasions repetitive spike potentials, indistinguishable from fibrillation, were seen with the muscle apparently at rest, but on each occasion these discharges disappeared when the patient was urged to relax, and it is clear that these

TABLE II

THE RESULTS OF FREQUENCY ANALYSIS OF THE ELECTROMYOGRAM IN PATIENTS WITH MYOPATHY

	Muscle Examined	No. of Insertions	Type of Spectrum Obtained		
			Shift	Broad Spectrum	Normal
Pseudohypertrophic Muscular Dystrophy (43 Cases)	Deltoid	6	2	3	1
	Biceps brachii	17	12	5	—
	Pectoralis major	16	12	4	—
	Supraspinatus	5	3	1	1
	Triceps	5	3	1	1
	Brachioradialis	3	1	2	—
	Quadriceps femoris	19	13	4	2
	Tibialis anterior	17	11	5	1
	Gastrocnemius	18	5	8	5
Facio-scapulo-humeral Muscular Dystrophy (12 Cases)	Pectoralis major	7	6	1	—
	Supraspinatus	5	1	3	1
	Biceps brachii	4	3	1	—
	Brachioradialis	3	2	1	—
	Orbicularis oculi	2	—	2	—
	Orbicularis oris	2	—	2	—
	Quadriceps femoris	6	4	2	—
	Gluteus maximus	4	2	2	—
Late Juvenile Type of Muscular Dystrophy (9 Cases)	Pectoralis major	12	10	2	—
	Deltoid	6	3	3	—
	Biceps brachii	14	11	3	—
	Supraspinatus	4	1	3	—
	Quadriceps femoris	7	4	3	—
	Gluteus maximus	2	1	1	—
	Tibialis anterior	7	4	3	—
	Gastrocnemius	3	—	1	2
Dystrophia Myotonica (15 Cases)	Flexor digitorum	10	8	2	—
	Thenar eminence	7	4	—	3
	Extensor digitorum	7	6	—	1
	Orbicularis oris	3	1	2	—
	Quadriceps femoris	7	2	3	2
	Tibialis anterior	12	11	1	—
	Gastrocnemius	8	3	5	—
Nevin Type of Muscular Dystrophy (3 Cases)	Pectoralis major	2	1	1	—
	Supraspinatus	2	—	1	1
	Biceps brachii	2	2	—	—
	Gluteus maximus	1	1	—	—
	Quadriceps femoris	2	1	1	—
	Tibialis anterior	1	1	—	—
Ocular Myopathy	Orbicularis oculi	1	1	—	—
	Biceps brachii	1	—	1	—
	Tibialis anterior	1	—	1	—
Distal Myopathy (Gowers)	Extensor digitorum	1	—	1	—
	Thenar eminence	1	1	—	—
	Adductor pollicis	1	1	—	—
	Tibialis anterior	1	1	—	—
Atypical Myopathy (6 Cases)	Pectoralis major	2	1	1	—
	Deltoid	2	2	—	—
	Biceps brachii	3	2	1	—
	Gluteus maximus	2	—	1	1
	Quadriceps femoris	4	3	1	—
	Tibialis anterior	4	2	2	—
	Gastrocnemius	3	1	2	—

were myopathic short duration potentials which were occurring because of incomplete relaxation of the muscle. It is important to recognize this pitfall which may give rise to considerable diagnostic difficulty. Single motor unit or fasciculation potentials which commonly occur spontaneously in motor neuron disease were not seen in any case of myopathy nor did any case show the characteristic grouped potentials of nerve irritation.

Spontaneous chains of oscillations of high frequency, attributable to muscular hyperexcitability, give rise to one of the most dramatic phenomena encountered in clinical electromyography. The characteristic "dive-bomber" or "quasi-musical" sound produced in the loudspeaker by this type of discharge is quite unmistakable (Buchthal and Clemmesen, 1941). This phenomenon is almost invariable in the myotonias on stimulation of the resting muscle with the needle point, but it has also been noted infrequently in progressive muscular atrophy, fatigued normal muscle, cramp, and anterior poliomyelitis (Richardson, 1951a, 1952). In the present series spontaneous activity of this type was recorded in all 15 cases of dystrophia myotonica and in six cases of myotonia congenita. It was also obtained, however, in 16 cases of muscular dystrophy without myotonia (11 pseudo-hypertrophic, two facio-scapulo-humeral, three late juvenile), in two out of 13 cases of progressive muscular atrophy, and in one case of dermatomyositis. The discharge was invariably less dramatic and less easily obtained in the cases of simple dystrophy than in those with myotonia, and as a rule the sound decreased in pitch much more slowly. It is of interest that a similar phenomenon, giving a "rasping" note slowly decreasing in pitch, was obtained on recording from shoulder girdle muscles during voluntary contraction in nine cases of muscular dystrophy. It is convenient at this point to mention the pattern of analysis produced by this phenomenon. As Richardson (1952) has mentioned, the individual components of the chain of oscillations sometimes have the dimensions of single fibre potentials, some resemble motor units, and others are polyphasic and bizarre. As would be expected, therefore, the spectrometer showed a broad spectrum type of response during recording of this activity; it was also noted that as the sound in the loudspeaker waned, those vertical lines of the spectrometer histogram corresponding to the higher frequencies fell out in turn, rather like a row of chorus girls falling one by one on to the stage. This "chorus girls" effect is amusing and, in the author's experience, it is quite diagnostic of the type of discharge under consideration, but it

cannot be considered important, as the electromyographic pattern is itself characteristic. It can be concluded that whereas dramatic, repetitive, and easily obtainable chains of oscillations of high frequency with the concomitant " dive-bomber " note remain characteristic of myotonia, similar but less impressive discharges may be obtained in patients with muscular dystrophy both at rest and on volition and in a number of other conditions ; the exact significance of these discharges remains obscure.

Volitional Activity.—Apart from the characteristic pattern of analysis already described, few observations of importance on the electromyographic pattern during voluntary contraction have been made in patients of this series. It has been possible to confirm Kugelberg's (1949) observations that even in grossly wasted muscles, with scarcely a flicker of voluntary contraction visible clinically, a sustained though highly polyphasic interference pattern may be obtained. It has also been confirmed that the amplitude of the action potentials of dystrophic muscle is greatly reduced in comparison with normal motor unit potentials, as would be expected in view of unit disintegration. These findings suggest that the muscular power represented by the dystrophic short duration potential is greatly reduced when compared with that of a normal motor unit potential.

It has also been noted that in recording from a muscle showing pseudohypertrophy, say the gastrocnemius, many parts of the muscle bulk may be electrically silent even during a powerful contraction ; presumably this observation can be correlated with the pathological finding of extensive fatty infiltration in such muscles.

Other Conditions.—The diagnoses of the patients examined in this section of the investigation have been noted in Table I. In this section attention will be confined to the pattern of analysis obtained in these cases, since this study is not concerned with the other electromyographic findings in these many disorders. In Table III the type of analysis obtained during voluntary contraction of stated muscles in these patients is given.

TABLE III

FREQUENCY ANALYSIS IN PATIENTS WITH OTHER NEUROLOGICAL DISORDERS

	Muscle Examined	No. of Insertions	Type of Analysis Obtained		
			Shift	Broad Spectrum	Normal
Motor Neuron Disease (13 Cases)	Pectoralis major	3	—	—	3
	Deltoid	6	—	1	5
	Biceps	2	—	—	2
	Brachioradialis	2	—	—	2
	Adductor pollicis	3	—	1	2
	Thenar eminence	3	—	2	1
	Quadriceps femoris	7	—	—	7
	Tibialis anterior	4	—	—	4
	Gastrocnemius	2	—	—	2
Polyneuritis (4 Cases)	Extensor digitorum	3	—	1	2
	Thenar eminence	3	—	2	1
	4th dorsal interosseus	2	—	—	2
	Tibialis anterior	5	—	—	5
Neuralgic Amyotrophy (2 Cases)	Supraspinatus	2	—	—	2
	Deltoid	2	—	—	2
	Triceps	1	—	—	1
Cervical Spondylosis (3 Cases)	Deltoid	2	—	—	2
	Thenar eminence	3	—	2	1
	4th dorsal interosseus	3	—	2	1
Myotonia Congenita (6 Cases)	Flexor digitorum	3	—	—	3
	Extensor digitorum	3	—	—	3
	Tibialis anterior	5	—	—	5
Amyotonia Congenita (1 Case)	Biceps brachii	2	—	—	2
Dermato-Myositis (2 Cases)	Quadriceps femoris	2	2	—	—
	Deltoid	2	1	1	—
Peripheral Nerve Injury (8 cases)	Serratus anterior	2	1	—	1
	Supraspinatus	2	—	1	1
	Thenar eminence	2	1	1	—
	4th dorsal interosseus	6	2	3	1
	Adductor pollicis	4	1	3	—
Facial Palsy (9 Cases)	Orbicularis oculi	9	1	7	1
	Orbicularis oris	9	2	7	—

It will be seen from the results outlined in Table III that in general the pattern of analysis seen in patients with neuropathy has been entirely different from that obtained in the myopathies. A shift of the spectrum to the right occurred only in the cases of dermatomyositis and in patients with regenerating peripheral nerve injuries or recovering facial palsy. From the principles already outlined these findings would be expected, since in dermatomyositis the disease process occurs within the muscle itself, as in myopathy, and as already mentioned, profuse polyphasic " recovery " potentials occur during recovery from a peripheral nerve injury. In all the patients with neuropathy, save one case of polyneuritis, the pattern of analysis for the particular muscles examined was normal, even after extensive exploration ; in one patient with polyneuritis (in

association with porphyria) an excessive amount of high frequency discharge was obtained from the forearm musculature, but the proportion was never sufficiently great in this case to produce a shift of the dominant frequency.

It is unlikely that clinical confusion will ever arise between cases of recovering peripheral nerve injury and myopathy; hence it can be suggested with confidence that this method provides a diagnostic tool for the unequivocal differentiation of myopathy and neuropathy. Both cases of dermatomyositis examined were referred for examination as cases of muscular dystrophy, and the correct diagnosis was reached only through the observation of an unusual pattern of muscular weakness combined with induration of subcutaneous tissue and minimal scleroderma-like changes in the skin of the hands and face. Clearly occasional cases of this condition will continue to cause confusion with muscular dystrophy; as would be expected from the physiological principles, electromyography is of no value in distinguishing these two conditions which must be separated by clinical and pathological methods.

Conclusions

The use of the audio-frequency spectrometer in the analysis of the electromyographic interference pattern has given results in patients with myopathy which are significantly different from those obtained in control subjects and in patients with other neurological disorders, with the exception of dermatomyositis and recovering nerve injury. It is suggested that this refinement of electromyographic technique is of considerable value in distinguishing between neuropathy and myopathy. The apparatus utilized in the investigation is probably too complex and expensive for routine use, but the results suggest that a simple type of analyser, designed on similar lines, is likely to be a valuable adjunct in diagnostic electromyography.

Summary

A method of frequency analysis of the electromyogram, utilizing an audio-frequency spectrometer, has been applied to the examination of 100 cases of myopathy, 26 normal subjects and 48 patients with other neurological disorders. The method was developed since electromyography has not given uniformly successful results in the diagnosis of myopathy, even though disintegration of the motor unit potential, with numerous polyphasic and short duration potentials, has been recognized in such cases. Analysis of the interference pattern of muscular contraction in normal subjects revealed a consistently normal pattern with the dominant response at 100–250 c.p.s., tailing off to zero at 800 c.p.s., save in facial muscles and small muscles of the hand, where a proportion of higher frequencies up to 1,250 c.p.s. was added.

In 98 of the 100 cases of myopathy the analysis obtained on contraction of at least one of the muscles examined showed a shift to the right of the dominant frequency compared with the pattern observed in normal subjects. These findings were most readily obtained from the superficial part of the muscle and in muscles severely affected by the disease process (pectoralis major, biceps brachii, quadriceps femoris, and tibialis anterior in the "proximal" types of muscular dystrophy, the long flexors and extensors of the fingers and tibialis anterior in dystrophia myotonica and distal myopathy). A similar shift of the dominant frequency occurred in patients with dermatomyositis and recovering peripheral nerve injury, but on extensive exploration in cases of neuropathy, including 13 patients with motor neuron disease, a normal analysis was practically invariable and no frequency shift was observed.

Spontaneous fibrillation was not observed in any case of myopathy; chains of oscillations of high frequency were recorded on probing in resting muscle in 15 out of 15 cases of dystrophia myotonica, six out of six cases of myotonia congenita, and in 16 out of 85 cases of muscular dystrophy with no clinical evidence of myotonia.

It is suggested that the addition of a method of frequency analysis as described enhances the value of electromyography in distinguishing between myopathic and neuropathic disorders.

I am grateful to Professor F. J. Nattrass for permission to publish this work and for his encouragement. The investigation could not have been carried out but for the kind cooperation of the Engineer-in-Chief, the Post Office Research Station, Dollis Hill, who lent the audio-frequency spectrometer. I am also indebted to Dr. G. A. Smart for his advice and to Mr. J. W. Osselton, Mr. Norman Clark, and Mrs. J. Molyneux for technical assistance. Diagrams and photographs are reproduced by permission of Mr. C. J. Duncan, of the Department of Photography, King's College.

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THORACIC INTERVERTEBRAL DISC PROLAPSE WITH SPINAL CORD COMPRESSION

BY

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The serious disorders of nervous function which may result from central protrusions of the intervertebral disc are exemplified in the thoracic region of the spine where this type of prolapse has acquired a sinister reputation, and the few cases treated by operation that have so far been reported bear witness to the severe damage that these protrusions inflict on the spinal cord and to the grave hazards attending their surgical removal. Thus, of Müller's (1951) four cases, three were left after operation with a complete or almost complete transection of the cord, and the fourth after a slight temporary improvement also developed a total paraplegia. Of Hawk's (1936) three patients who survived operation, all with severe pre-operative spinal cord damage, one was made worse by, and the other two showed no benefit from, surgery. Mixter and Barr (1934) reported three cases, two of which developed a total transverse lesion and the third eventually showed slight improvement. So of 10 cases of central thoracic disc prolapse treated surgically, six were left with virtually complete cord transection and there was little or no worthwhile improvement in the other four, a unique morbidity rate among benign compression lesions of the cord.

It is my purpose to describe a series of 11 cases all proven by operation which does to some extent confirm this grave picture, but also reveals that it is possible to make an accurate pre-operative diagnosis of this type of herniation and to remove a certain number of these protruding discs not only without further damage to the spinal cord but with almost complete restitution of neurological function.

Incidence

These 11 cases have appeared in a modest total of 250 disc protrusions at all levels, an incidence of 4% for thoracic prolapse, which is a greatly exaggerated one owing to the restricted choice of cases for admission; the true proportion is probably in the neighbourhood of two or three per 1,000 disc prolapses as reckoned by Love and Kiefer (1950) in the cases at the Mayo Clinic. It is of interest that among these 250 cases there were also 14 patients with central lumbar disc protrusions producing cauda equina compression and 15 central cervical protrusions with spinal cord involvement.

Site

There are 12 thoracic intervertebral discs, the first lying between the first and second thoracic vertebrae and the twelfth between the last thoracic and first lumbar vertebrae, and the portion of the spinal cord which is exposed to compression by central protrusions of these discs extends from the third thoracic to the fifth lumbar spinal segment.

Fig. 1 shows the disc levels at which the protrusions

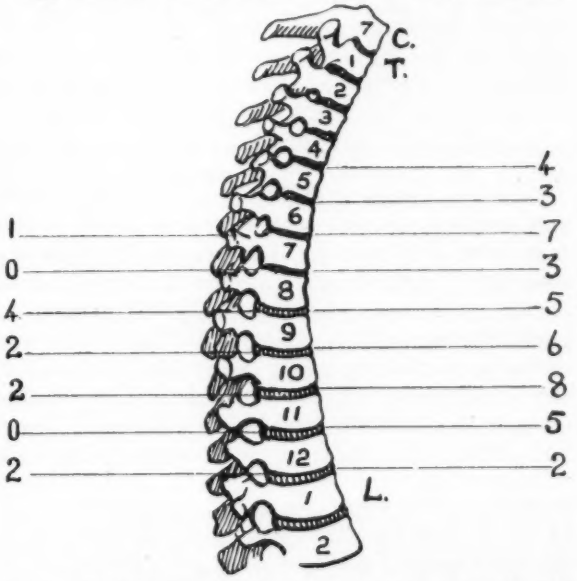

FIG. 1.—The figures on the left refer to the 11 cases in this series. The list of 43 on the right includes an additional 32 reported cases: Bradford and Spurling (1945) one; Elsberg (1931) two; Hawk (1936) four; Love and Kiefer (1950) 17, including three of lateral disc protrusion; Mixter and Barr (1934) four cases; and Müller (1951) four cases.

have occurred. The list on the left of the diagram refers to this series of 11 cases. To obtain a more accurate picture an additional 32 reported cases, verified by operation or necropsy, have been collected making a combined total of 43, which is shown in the list to the right of the diagram. This demonstrates that central protrusions are confined to the lower nine intervertebral discs without, however, a predilection for any two of them as pertains in the lumbar and cervical regions.

Sex, Age, and Trauma

Table I shows that there are nine men and two women in the series, in accord with the general rule that disc protrusions at any level tend to be more common in males. This predominance is confirmed, but on a lesser scale, by the larger group of 43 cases in which the ratio is as five to three. It is also apparent from this table that thoracic disc prolapse is a disease of middle and late adult life, the youngest patient being 38 years old and the average age of the whole series 50 years.

TABLE I
SEX, AGE, AND INCIDENCE OF TRAUMA IN 11 CASES

Case No.	Sex	Age	History of Trauma
1	Male	48	Yes
2	Male	44	No
3	Male	38	Yes
4	Male	52	No
5	Male	62	No
6	Female	44	No
7	Female	63	Yes
8	Male	54	No
9	Male	61	No
10	Male	44	No
11	Male	45	No

Direct or indirect trauma plays little part in the aetiology of prolapse in this region of the spine. In only three of these patients was there any possible association with injury. In one patient (Case 1) the first symptom of back pain, which preceded the onset of neurological signs by 10 years, appeared while he was lifting some heavy weights. In the second (Case 3) back pain appeared a few days after an episode of vigorous gardening and was followed five months later by cord compression. The third patient (Case 7) twisted her back while turning suddenly and her first symptoms appeared next day. In the remaining eight cases there was no history of injury or unusual exertion and the patients were otherwise in normal health.

Clinical Features

A description of the neurological findings in each of these patients appears in some detail in the case histories at the end of this paper. A study of them confirms what might have been expected: that there are no symptoms or signs which are characteristic of compression of the spinal cord by a thoracic disc prolapse and that they do not differ materially from those produced by spinal neoplasms. The following case illustrates some of the common clinical features of the condition and also the pitfalls that may beset the diagnosis in the early stages.

Case 11.—A man aged 45, a research chemist, seven months before admission developed pain in his back at about the dorsi-lumbar junction, slightly to the left of the midline. It was intermittent, had an aching character, and was made worse when lying down, but was unaffected by coughing and straining. He had sustained no injury to his spine.

A few days later numbness appeared in the right thigh and quickly spread down the leg to the toes and upwards to the level of the umbilicus. Two weeks later the right leg became stiff, weak, and unreliable so that he tended to fall. He was investigated at another hospital two months after the onset of symptoms and was found to have a spastic right leg with increased reflexes in both legs and a right extensor plantar response. There was patchy impairment of sensation to pain and light touch over the right leg and the inner side of the left ankle. Lumbar puncture at that time showed no evidence of spinal block and the fluid contained three cells and 25 mg. % of protein. He was thought to be suffering from atypical disseminated sclerosis and was sent home. His symptoms progressed steadily and five months after the onset weakness and numbness appeared in the left leg and he also developed urgency of micturition and difficulty in defaecation.

Neurological Examination.—There was a little weakness in the lower abdominal muscles and considerable weakness of both legs with increased tone, more marked on the right, and sustained ankle clonus. Coordination was grossly impaired.

The abdominal reflexes were absent. The knee and ankle jerks were exaggerated, more so on the right. Both plantar responses were extensor.

There was a level at the seventh thoracic dermatome on both sides below which there was some impairment of pain sense and a more definite level at the tenth dermatome below which there was considerable impairment to pain, light touch, and temperature, but not complete loss. Position sense was absent at the toes and reduced at the ankles. Vibration sense was absent over the right tibia and reduced on the left.

He was just able to walk without support, but very unsteadily on a wide base.

He was later proved to have a prolapse from the ninth-tenth intervertebral disc. (The details of investigation are described at the end of this paper.)

In general the symptoms of cord compression appeared in the chronological order of sensory

(pain, tingling, burning, coldness, or numbness), motor, and sphincter, and once begun the course was that of a relentless and fairly rapid progression so that in eight of the 11 cases the time elapsing from the first symptom of compression to the development of a marked neurological deficit warranting surgery was less than seven months. As exceptions to this rule there were three cases (Nos. 6, 4, and 7) with histories of 14 months, two years, and 10 years respectively, and in the last of these the course did show some remittent features.

Among the first group of eight cases there were three (Cases 5, 8, 9), and these will be referred to again when dealing with the mechanics of compression, in which the symptoms of a severe paraplegia developed acutely in the course of several days, but in only one (Case 9) was a recent soft disc prolapse found at operation. In the other two cases the protrusion was bony hard and had obviously been present for a considerable time, probably years, and the sudden appearance of a paraplegia was thought to be due to interference with the blood supply to the cord.

In most of the patients the symptoms started in both legs at the same time, or if they did appear first in one leg the other was soon involved, so that by the time of admission to hospital neurological examination revealed signs indicative of compression of both halves of the spinal cord. In three patients, however (Cases 1, 2, 3), the presenting neurological picture was that of a Brown-Séquard syndrome, resulting from compression of one half of the spinal cord by a prolapse situated just to one side of the midline of the spinal canal. That protruded cervical discs could cause either bilateral or strictly unilateral cord compression was noted some years ago by Stookey (1928) and in practice this distinction is not as artificial as it may at first appear. This observation of Stookey's also holds true for the thoracic region, but here the hemicompression syndromes are proportionately less frequent and not so clearly defined as with cervical protrusions because of the larger size of the thoracic prolapse relative to the smaller bulk of the spinal cord in this region. In the three representative cases mentioned above the syndromes are by no means complete.

In the sensory field, as would be expected with an anteriorly situated lesion, pain and temperature sense were more affected than light touch and the modalities subserved by the posterior columns ; in fact in some cases the latter structures were practically uninvolved.

One patient (Case 11) showed sensory impairment extending up to a dermatome level considerably higher (four segments) than that of the spinal segment actually under compression and gave rise to some confusion in localization. This phenomenon has been previously noted in thoracic prolapse (Hawk, 1936) and is of course well recognized, although not satisfactorily explained, in association with spinal neoplasm and in cervical disc prolapse.

Sphincter disturbance was not a prominent symptom and, except for one case, when it did occur it appeared late in the course of the compression. In six patients there were no bladder or rectal disturbances whatsoever, often despite severe weakness of the legs. In three cases there was mild urgency or dribbling on micturition, and in only two cases, both with gross paraplegia, was there loss of sphincter control (Cases 7 and 10).

Pain

A feature of central disc prolapse which is common to the cervical and thoracic regions is the inconstant association of pain, either local or radicular. In nearly half of these cases (five out of 11) there was no complaint of pain at any time in the evolution of the clinical picture. Of the six patients who did experience pain, four (Cases 1, 3, 9, and 11) felt it locally in the back, where it preceded the onset of cord symptoms by a variable time ranging in the individual case from a few hours to 10 years. The pain was felt in the midline of the spine, sometimes extending a little to one side, and was located in the upper lumbar or thoraco-lumbar regions irrespective of the level of the prolapse. It was usually described as having an aching character, was not particularly severe, and often occurred in attacks similar to lumbar disc pain. In the remaining two patients (Cases 5 and 8) the prolapse was at the D.12/L.1 level and nerve root pain was produced which had a characteristic distribution extending from the upper lumbar region down the front and back of each lower limb as far as the ankle, but predominating in the anterior aspect of the thighs. This radiation was explained by the location of the prolapse where it was anatomically possible for it to involve the majority of the lumbar and also the upper sacral nerve roots as they lie closely grouped together round the conus medullaris.

Investigations

Changes in the Cerebrospinal Fluid.—Table II illustrates the degree of block on manometry and the protein content of the cerebrospinal fluid in the 11 cases. Five of the patients had a complete or almost complete spinal manometric block with, in the four cases in which it was recorded, considerable

TABLE II

MANOMETRIC CHANGES AND PROTEIN CONTENT OF CEREBROSPINAL FLUID

Case No.	Manometric Results	Protein Content of C.S.F. (mg. per 100 ml.)
1	No block	55
2	No block	Not recorded
3	Partial block	30
4	Complete block	Yellow fluid; protein content not recorded
5	Complete block	Yellow fluid 175
6	Complete block	95
7	Complete block	150
8	Almost complete block	200
9	Partial block	100
10	Partial block	20
11	No block	50

elevation of the protein content of the cerebrospinal fluid to 200, 175, 150, and 95 mg. per 100 ml. respectively. A partial block or "sticky" manometrics was recorded in three patients (Cases 9, 3, and 10) with a protein content of 100, 20, and 30 mg. per 100 ml. In the remaining three cases there was no evidence of block and the cerebrospinal fluid protein was 55 and 50 mg. per 100 ml. respectively in the two cases in which it was recorded.

It should also be noted that in two of the patients (Cases 4 and 11) both the cerebrospinal fluid and the hydro-dynamics were completely normal at a time when considerable neurological damage was present. The former patient went on to develop a total spinal block a year later and removal of the disc at that stage aggravated the paraplegia.

Perhaps the important deduction from these findings is not that thoracic disc prolapse can cause a complete spinal block but that in the early stages of the condition the manometrics and cerebrospinal fluid may be entirely normal, and even in the later stages of gross neurological disability there may be at most a partial block or a slight increase in the protein content.

Radiology.—In contrast to the clinical features of prolapsed thoracic discs, which are of little value in positive diagnosis, some of the radiographic changes form a reliable guide, and it should be possible by this means to make a pre-operative diagnosis in the majority of cases.

Arthritic lipping of the vertebral bodies in the thoracic spine, and often in the lumbar region as well, is frequently seen in these patients but appears to be no more common or more extensive than that observed in patients of this age group without disc prolapse. Narrowing of the affected disc was not a constant feature and in fact was often seen in discs which had not protruded.

With regard to calcification of the nucleus pulposus, however, it is an entirely different story. Of these 11 cases of thoracic prolapse, no less than eight showed unequivocal calcification in a nucleus pulposus. The significance of this finding was brought out by a study of the radiographs of the thoracic spine of 100 cases of similar age group who had had radiographs taken because of local spinal symptoms resulting from injury, arthritis, etc., and in which nuclear calcification was observed in only four instances compared with an incidence of over 70% with disc prolapse.

Again, in 25 patients suffering from thoracic spinal compression by neoplasms such as neurofibromata, meningiomata, and metastasic tumours, drawn from a similar age group, there were no instances of nuclear calcification.

The calcified disc, however, is not necessarily strictly related to the level of the prolapse. Table III shows that in five of these eight patients the calcification occurred only in the nucleus pulposus of the disc which had actually herniated. In two other patients there were several calcified discs (Cases 2 and 4 respectively) of which one in each case corresponded to the site of the prolapse. In the final case the calcification was present in a single nucleus but this was situated four disc spaces above the protrusion. It would seem that nuclear calcification in the thoracic region is indicative of a degenerative change of such a nature as to render the disc liable to prolapse, but this change may also be present in other discs as yet uncalcified, and it may happen that prolapse occurs from one of the latter.

TABLE III

INTERVERTEBRAL DISC LEVEL OF CALCIFICATION, RELATIONSHIP TO THE SITE OF PROLAPSE, AND MYELOGRAPHIC APPEARANCES

Case No.	Level of Disc Prolapse	Level of Calcification	Myelographic Appearances
1	D 9/10	D 9/10	Oval defect 2×1·5 cm.
2	D 8/9	D 8/9	Partial block
3	D 8/9	D 8/9 and 9/10	Oval filling defect
4	D 6/7	D 6/7	Complete block
5	D 12/1	D 8/9	Complete block
6	D 8/9	D 8/9	Circular filling defect 1·5 cm. diam.
7	D 10/11	No calcification	Partial block
8	D 12/1	No calcification	Complete block
9	D 10/11	No calcification	Complete block
10	D 8/9	D 8/9	—
		D 9/10	—
		D 10/11	—
		D 11/12	Partial block
11	D 9/10	D 9/10	Oval filling defect 2×1·5 cm.

In a person suffering from spinal cord compression in the thoracic region calcification of a nucleus pulposus is an important finding, and as a general guide it may be stated that if the segmental level of the lesion corresponds with a calcified

nucleus then the diagnosis of a protruded disc is practically certain, and even when the segmental level does not correspond a herniation is still the most likely diagnosis.

The calcification is best seen in the lateral view. Its appearance in the nucleus has no specific features and, as Figs. 2, 3, and 4 demonstrate, it varies considerably. It may consist of a uniformly dense plaque which occupies part or the whole of the nucleus (Fig. 2) or it may be arranged in the form of flakes or granules (Figs. 3 and 4).

At operation the periphery of some of the protrusions was found to be quite heavily calcified, but it has been possible in only one (Case 6 and Fig. 5) to demonstrate this circular rim by radiography. In the others the shadow was presumably too faint to be discerned amid the dense bone in the vicinity.

Myelography.—The thoracic region is notoriously difficult to screen, partly owing to the superimposed shadows of the heart and great vessels which tend to obscure detail, and partly owing to the convexity of this portion of the spinal column, which is accentuated in these older patients. As soon as the contrast material runs over the thoracic hump whether going up or down the spinal canal it tends to glide rapidly over the remaining vertebrae to gain the lordosis of the lumbar or cervical regions, often breaking up into globules as it does so, and during this rapid flow it is possible to overlook the small filling defects which may be the only sign of a thoracic prolapse.

Table III shows that on myelographic screening of these 11 cases four showed a complete block which often had a convex border to it (Fig. 6 and Case 9). Of the remaining seven, three revealed a partial block without characteristic features and in four the only evidence of a disc was a filling defect in the column of contrast material. This filling defect has quite typical features (Fig. 7). It is circular or oval in shape, varying from 1·5 to 2 cm. in diameter, and overlies the intervertebral disc space and adjacent portions of the vertebral bodies. If good lateral views can be obtained (and this is often very difficult) it is usually possible to demonstrate that the obstruction lies anteriorly.

In two of the patients (Cases 3 and 8) in the lateral view of the myelogram a " double profile " could be seen at the level of the affected disc (Fig. 8), an appearance which depends on the prolapse being situated to one side of the midline so that part of the contrast medium runs over the convexity of the prolapse and part runs in the dural gutter alongside it, giving two levels which may be as much as a centimetre apart.

Mechanics of Spinal Cord Compression

It is pertinent at this stage to enquire briefly into the means whereby the herniated disc produces interference with cord function. In some cases there is manifestly a true compression as a result of the large size of the protrusion which may occupy the greater part of the diameter of the spinal canal and squeeze the cord backwards against the lamina and ligamentum flavum. In other cases, however, the prolapse may be relatively small and incapable of " compressing " the cord as a whole, and another explanation has to be sought. This problem has previously arisen with regard to cervical disc lesions, and two explanations have been put forward. One concerns the ligamenta denticulata and the other the blood supply to the spinal cord.

Kahn (1947) made the suggestion that the ligamenta denticulata resisted the backward displacement of the cord and produced traction on, and distortion of, the nerve fibres in the vicinity of the attachment of the ligaments to the transverse meridian of the cord. This may well be the explanation of the neurological signs in some of the cervical disc compressions, particularly in those cases which present with almost exclusive involvement of the pyramidal tracts.

In some cases of thoracic (and cervical) disc compressions, however, it has been proposed that interference with the blood supply is the basis of the pathological change, in particular as a result of obstruction or occlusion of the main arterial trunk—the anterior spinal artery. This would best explain (1) the appearance of a neurological picture resembling that which occurs in known cases of anterior spinal arterial thrombosis ; (2) the abrupt onset in some patients of extensive damage to the cord when at operation a hard prolapse evidently of very long standing is found; (3) the occurrence of degenerative changes amounting to necrosis of the cord substance ; and (4) the severe damage inflicted on the cord out of all proportion to the size of the protrusion, and the poor recovery that may result despite the complete removal of the prolapse.

It was therefore with the hope that some light would be thrown on this problem that the spinal cord in the one patient coming to necropsy (Case 6) was examined, but it was clearly shown in this case that there were no demonstrable changes in the main vessel which could be invoked as the cause of the neurological picture or of the post-operative transverse lesion.

The patient had a 14-month history of progressive paraplegia and developed a total transverse

FIG. 2.—Lateral radiograph of the thoracic spine in Case 10 showing the dense calcification in the nucleus pulposus of three of the four calcified discs.

FIG. 4.—Lateral radiograph of Case 6 showing the granular appearance of the calcification in the nucleus of the eighth-ninth intervertebral disc.

FIG. 3.—Lateral radiograph of Case 11. Flaky calcification is present in the nucleus pulposus of the ninth-tenth intervertebral disc.

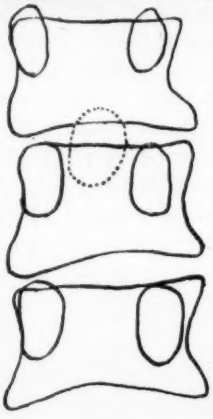

FIG. 5.—Antero-posterior radiograph of Case 6 to show the ring of calcification in the wall of the prolapse from the ninth-tenth intervertebral disc.

FIG. 6.—Myelogram of Case 9 illustrating the complete obstruction just below the tenth-eleventh disc with a convex lower border. It was similarly outlined from above.

lesion after operation, succumbing nine months later. The spinal cord, dura, and prolapse were removed in one piece and then sectioned en bloc (Fig. 9). The extreme distortion of the fibre tracts is well shown but the anterior spinal artery, although in close apposition to the prolapse, is quite patent without any thickening of its wall, and this appearance was confirmed at higher and lower levels. The anterior spinal vein also showed no evidence of damage.

Surgical Results

The results of surgery in these cases are not such as to foster complacency. There were in all five bad results (Cases 2, 4, 5, 6, and 7) and two of these patients died.

Of these five patients, three (Cases 5, 6, and 7), all severely disabled before operation, developed a total transection post-operatively. One (Case 5) died four months later from urinary sepsis without having made any neurological improvement. Case 6 showed slight recovery in motor power and pain sensation and then died nine months after operation from urinary sepsis and bed-sores. The third patient (Case 7), who showed gross degenerative changes in the spinal cord at operation, has made no recovery in four years. The prolapse was excised completely in the first case but in the other two surgical intervention was confined to a decompression.

FIG. 7.—Myelogram of Case 11 showing the typical oval filling defect at the level of the calcified disc (D 9/10).

FIG. 8.—Lateral radiograph of the myelogram in Case 3. Calcification is present at the eighth-ninth and ninth-tenth disc levels. The double profile of the contrast medium outlines the prolapse which has occurred from the upper, less calcified, disc.

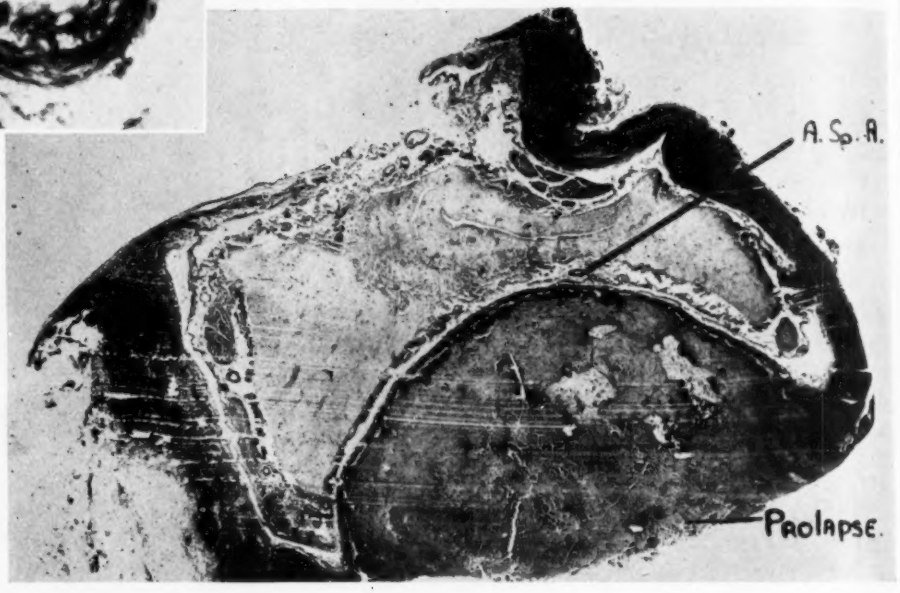

FIG. 9.—Section through the prolapse, dura mater, and spinal cord in Case 6.
The distortion of the cord and its close apposition to the surface of the prolapse is well shown. The anterior spinal artery although small (as is usually the case at this level) is quite patent and its wall shows little pathological change. (Inset) Magnified view of the artery. × 160.

series, had a complete manometric spinal block and a total, or almost total, myelographic block indicating an unusually large protrusion. The third feature was the presence at operation of naked eye changes of focal degeneration in the substance of the spinal cord. This was seen in three of these five patients (the only examples in the series) and consisted of areas in the cord varying in diameter from a few millimetres to a centimetre or more, overlying the apex of the protrusion and presenting a translucent, waxy appearance with grey or blue discoloration.

It is apparent that the unfortunate results of surgery in these five patients were to a great extent predetermined by the severe structural damage to the spinal cord, as manifested by the severity of the neurological disability, the large size of the protrusion, and visible degenerative changes.

Two patients (Cases 2 and 4), both showing macroscopical degenerative changes in the cord substance at operation, and the latter with a severe pre-operative paraplegia, were rendered worse by removal of the protrusion and then made slow, incomplete recoveries over several years so that they were eventually able to get about with the aid of sticks. Six years after operation Case 4 again retrogressed to an almost total paraplegia, possibly from further protrusion of the disc.

Study of these five cases reveals that in addition to their tragic outcome they had several other features in common. The first of these was the severity of the neurological disability before operation ; and four of these five patients had gross paraplegias at the time of admission to hospital. The second feature was the size of the compressing lesion, and again four of these cases, out of five in the whole

The remaining six patients present a happier picture.

Case 8.—This man had a severe paraplegia so that he was able to walk only a few steps and then with severe pain in the back and legs. Sensation was less affected. Surgery was limited to a decompression as the protrusion was of bony hardness. This procedure seems to have arrested the progress of the condition and there has been slight improvement in so far that he is able to get about his house in reasonable comfort and can walk distances up to 200 yards with a stick and stand for periods up to half-an-hour. Sphincter control is normal.

Case 1.—In this case there was evidence of a fairly severe Brown-Séquard syndrome. Most of the disc prolapse was excised ; this temporarily exacerbated the

physical signs, but in the 12 months since operation he has made steady improvement. He is now able to walk distances up to two miles and has returned to work driving a van. Bladder control is practically normal and his original back pain, present for 10 years, has disappeared.

Case 10.—This patient was grossly disabled and was unable to lift his legs against gravity. There was dense sensory loss and retention of urine and faeces. A complete removal of the prolapse was carried out. It is now two and a half years since operation. He is able to walk 500 yards with the aid of sticks and has returned to full-time sedentary work. Some urgency of micturition persists but bowel control is normal.

Case 11.—This man had a similar but slightly less severe neurological deficit. The major part of this prolapse was removed but the right hand margin was inaccessible. It is now three and a half years since operation and for the last two and a half he has been back at full work and can walk and run normally, and drives a car. His only residual symptom is that of a sensation of cramp and tightness in the muscles of the left leg and subjective numbness of the feet.

Case 9.—This patient showed considerable motor weakness with milder posterior column sensory impairment as a result of an acute prolapse of softened nucleus pulposus, which was completely excised. At 10 months after operation he can walk any distance with one stick. He has no pain or sensory change and sphincter control is normal.

Case 3.—This was the youngest patient in the series and presented with a mild Brown-Séquard syndrome. The disc was totally excised. It is now three years since operation and for the last two and a half of them he has been back at full work, and can walk, run, dance, and dig as well as he ever did. His remaining symptoms are those of a sensation of tightness in the calves and some loss of potency.

This last case represents the best result obtained in this series and is perhaps accounted for by the short history of compression (two and a half months), a small prolapse, and the absence of degenerative change in the cord at operation.

Surgical Management

The surgery of compressing lesions which lie anterior to the spinal cord is never easy and is made immeasurably more difficult in disc prolapse by the changes produced in the cord which render it vulnerable even to the slightest surgical trauma. In addition the anatomical relationship of the cord to the protrusion (Fig. 9) makes access extremely difficult. The consistency of the prolapse is sometimes another hazard, for it may be composed of a small central area of cartilage set between bony spurs which project from the vertebrae above and below (Case 7), and which cannot be chiselled away without risk of further injury to the nervous structures. However, a recent report by Allen (1952) suggests that it is possible to remove these spurs by the technique he describes without increasing the damage to the cord, and it may be that a more radical attitude will be taken in the future.

In the surgical approach the most important factor is an accurate pre-operative diagnosis and the knowledge from the start that one is dealing with a discal hernia. This knowledge should ensure removal of the lamina with the utmost caution and so prevent any further compression of the cord against the unyielding prolapse during the exposure. Other technical points are : that the laminectomy should be extensive consisting of at least three, preferably four, laminae ; in a narrow spinal canal I have no hesitation in removing the pedicle on the side of the maximum protrusion to gain a more lateral approach ; all the ligamenta denticulata in the operative field should be divided. It is perhaps hardly necessary to emphasize the extreme gentleness required in lifting up the edge of the spinal cord to gain access to the hernia, and for this reason it is best to remove the prolapse by the intradural route so that the minimal displacement of the cord that is permitted can be kept under direct control. The soft central area of the prolapse is usually easily removed but the hard rim may require to be excised piecemeal with a fine gouge.

Once the protrusion has been exposed a decision has to be made either to attempt its removal or to be content with a decompression with or without division of the ligamenta denticulata. If there is an area of necrosis in the cord, and this is a most ominous sign even when the associated neurological deficit is not severe (Case 2), a decompression is all that should be done. If the type of prolapse is that of the transverse bony spur it is again probably safer not to attempt a radical excision (but this view may require modification in the future). On the other hand, if the cord is not grossly abnormal and the prolapse has the more usual rounded, sometimes mushroom, shape it is imperative to remove it entirely for it is only by this means that any considerable recovery in neurological function can result.

It is evident that the reputation these central thoracic disc protrusions have acquired is not undeserved. It is only by early diagnosis before permanent changes occur in the spinal cord that an improvement in surgical results will be obtained, and this should be possible, if with a progressive thoracic spinal lesion the significance of a calcified nucleus pulposus is appreciated, and myelography with careful screening is undertaken at a stage when

the manometrics and cerebrospinal fluid may well be normal.

Case Histories

The *post-operative* condition of the 11 patients is described mainly in terms of recovery of function and the finer details of neurological examination are omitted.

The first three cases are examples of incomplete Brown-Séquard syndromes.

Case 1.—A man aged 48, a greengrocer, four months before admission developed numb feeling in both legs followed in a few days by weakness, particularly in the left leg, which progressed steadily. He had no sphincter disturbance. Ten years previously, after lifting a heavy weight pain appeared in the upper lumbar region and had occurred intermittently ever since. It had an aching character, did not radiate, and was not severe, usually being relieved by the application of heat. He had also contracted syphilis 21 years previously for which he had received treatment at the time.

Neurological Examination.—There was generalized weakness of the left leg with increased tone but no clonus, and impaired coordination.

Reflexes were exaggerated with an extensor plantar response. In the right leg no abnormality of power, tone, or reflexes was discerned.

On the left side sensation was normal except for the absence of vibration sense over the tibia. On the right there was a sharply defined level at the twelfth thoracic dermatome below which there was impairment of pain, temperature, and light touch. Vibration sense was normal. The sense of position and passive movement was unaffected in both legs.

The patient could walk unaided, rather hesitantly, circumducting the left leg.

Radiography.—Radiography showed dense mottled calcification of the ninth-tenth nucleus pulposus without narrowing of the disc space and arthritic lipping of D.11 and 12.

Myelography.—Myelography showed an oval filling defect 2 by 1·5 cm. at the level of the calcified disc.

Lumbar Puncture.—Manometrics were normal. The fluid contained one cell per c.mm. and 55 mg. of protein per 100 ml. The Wassermann reaction was negative, and the Lange reaction unchanged.

Operation.—Laminectomy of the ninth, tenth, and eleventh thoracic vertebrae was performed. A firm swelling was felt anteriorly through the dura. The spinal cord was kinked backwards by a hard swelling indenting the dura anteriorly, and was pale with some venous congestion. The dura and capsule of the prolapse were incised and soft, stringy disc tissue extruded. Complete removal, partly intra- and partly extra-dural, was effected.

Post-operative Course.—The patient developed retention of urine and impairment of pain, temperature, and light touch in the left leg. After 10 days the bladder recovered and power and sensation improved.

At one year after operation the patient can walk two miles with a stick and has returned to full work driving a van. Bladder control is practically normal. The original back pain has gone. Improvement still continues.

Case 2.—A man aged 44, a van driver, six months before admission developed a sensation of coldness and numbness on the outer side of the left leg below the knee, which had persisted but not extended. There had been weakness and stiffness of the right leg and similar changes in the left leg for six weeks, but no pain, no sphincter disturbance, and no spinal injury.

Neurological Examination.—There was moderate generalized weakness in the right leg and increased tone, with ankle and patella clonus. In the left leg power, tone, and coordination were normal.

Abdominal reflexes were absent. The right knee and ankle jerk were exaggerated. The left deep reflexes were normal. The plantar responses were extensor.

Sensory change was confined to diminution of pain sense on the outer side of the left leg from knee to ankle. All other modalities were normal.

Radiography.—Arthritic lipping of the bodies of most thoracic vertebrae was seen, and spotty calcification in the anterior portion of the nucleus pulposus of the eighth-ninth disc with narrowing of the disc space.

Myelography (Cisternal).—Contrast was partially arrested just above the eighth-ninth disc.

Lumbar Puncture.—This showed normal manometrics. Protein was not recorded.

Operation.—Laminectomy of the seventh, eighth, and ninth vertebrae was performed. A firm swelling was felt through the dura at D.8 level. The cord was bulged backwards by a small tense prolapse from the eighth-ninth disc. Just to the right of the midline at the convexity of the bulge there was an area in the cord substance, 3 mm. in diameter, which had a blue, translucent appearance. On incision of the prolapse soft, yellow tissue extruded. A complete intradural removal was effected.

Post-operative Course.—The patient had a complete flaccid paralysis of both legs. Below the eleventh thoracic dermatome all sensation was absent on the right but on the left pain and light touch could be felt although diminished. Urine and faeces were retained. Movement began to return in the feet in one month and there was slow recovery for 18 months after which his condition has remained stationary to the present time (11 years). He can stand, but with flexion of all joints, and can walk slowly with two sticks. He has dribbling incontinence and wears a rubber urinal all day and has a bottle in bed at night. The bowels act every fourth day with aperients. There is extreme spasticity of both legs with clonus and exaggerated reflexes and extensor plantar responses. Flexor spasms occur on the slightest

stimulus. There is a level at the eleventh thoracic dermatome below which pin prick is impaired and thermal sensation is absent. Light touch is well appreciated. Joint sense is reduced in the toes.

Case 3.—A man aged 38, a business executive, nine months before admission, a few days after some heavy work in his garden, developed an aching sensation to the left of the midline of the back at about the dorsi-lumbar junction, and this ache had persisted. For two and a half months he had been unsteady on his feet and for the same time the right lower limb had been numb from the hip downwards, most evident on the lateral aspect of the leg and foot, with a tingling sensation in the toes of both feet. There was no sphincter disturbance.

Neurological Examination.—Slight weakness of both legs was detected predominating in the left and more marked peripherally, with increased tone, ankle and patella clonus on the left and patella clonus on the right.

The knee and ankle jerks were very brisk, particularly on the left, and the plantar responses were extensor.

Sensory change was confined to the right side. Below the twelfth thoracic dermatome there was impairment of pain and temperature with sparing of sacral segments. Light touch was normal. Vibration sense was absent at both malleoli. Joint and position sense was unaffected.

Radiography.—Calcification in the nucleus pulposus of both the eighth-ninth and ninth-tenth discs was seen, more extensive in the latter, with slight narrowing of the eighth-ninth disc.

Myelography.—Myelography showed an oval filling defect at the eighth-ninth disc level. In the lateral views a " double profile " (Fig. 8) was seen.

Lumbar Puncture.—Manometrics showed a partial block with a " sticky " rise and fall to 250 mm. H_2O. The fluid contained 1 cell per c.mm. and 30 mg. of protein per 100 ml.

Operation.—Complete laminectomy of the eighth and ninth vertebrae was performed. A hard swelling was felt anteriorly. The spinal cord was displaced backwards by a spherical protrusion from the eighth-ninth disc lying mainly to the left of the midline. The spinal cord substance appeared normal. On incision through the dura and capsule soft, degenerate cartilage extruded. The harder rim was removed piecemeal.

Post-operative Course.—The legs were much weaker for a few days, but after that there was steady improvement. At three years after operation the patient can walk, run, dance, and dig as well as ever. Sensation is normal. His remaining symptoms are those of tightness in the calf muscles and some loss of potency.

Case 4.—A man aged 52, an insurance broker, two years before admission developed numbness in the left big toe which spread up the leg within a few months. Six months after the onset a similar change appeared in the right leg associated with weakness of this leg. After a further six months weakness started in the left leg. Both the motor and sensory loss progressed relentlessly so that at three months before admission a stage was reached where he was numb from the costal margin downwards and could take only a few faltering steps with support. Flexor spasms had been present for 12 months. Rectal control was normal, as was the initiation of micturition but he had difficulty in knowing when the bladder was empty. There had been no pain at any time. Six months after the onset of symptoms he had been investigated elsewhere when the cerebro-spinal fluid and manometrics revealed no abnormality and a myelogram did not show a block.

Neurological Examination.—Some loss of power in the abdominal muscles with gross loss in both legs was perceived but a small range of movement was possible at all joints. Tone was increased without clonus.

The abdominal reflexes were absent. The knee and ankle jerks were exaggerated on the left, normal on the right. Plantar responses were both extensor.

There was a level at the sixth thoracic dermatome below which pain was considerably impaired and temperature sense was lost. From the sixth to the tenth dermatome the skin was hyperaesthetic to light touch and below this hypoaesthetic. These sensory changes extended down to cover the whole of the right leg but on the left leg there was relative sparing below the third lumbar dermatome. Position sense was reduced at the toes. Vibration sense was diminished at all bony points in the legs.

Radiography.—Mottled calcification was seen in the nucleus of the sixth-seventh disc.

Cisternal Myelogram.—This showed a complete block just above the area of calcification.

Lumbar Puncture.—A complete block was demonstrated. The fluid was yellow with increased protein (precise amount not recorded).

Operation.—Laminectomy of the fifth, sixth, and seventh thoracic vertebrae was performed. A hard swelling was palpable anteriorly. The spinal cord was angled backwards by a smooth disc prolapse occupying two-thirds of the diameter of the spinal canal. In the cord substance there was an area, $\frac{1}{2}$ in. long and $\frac{1}{4}$ in. wide, of a dark purple lying just to the left of the midline. On incision of the dura and capsule yellow degenerate disc tissue extruded. Its hard rim was completely removed intradurally. A few drops of yellow fluid were aspirated from the discolored area of the cord.

Post-operative Course.—The patient had almost complete motor and sensory paralysis, with retention. A few degrees of movement remained in the toes and there was patchy retention of light touch over both legs. Recovery in motor power started six weeks later and continued slowly so that he could walk with sticks at 11 months. Maximum recovery was attained four years after operation when he could walk with one stick for 15 minutes, could drive a car, and had control of the bowel and bladder although some urgency of micturition persisted. At six years a further deterio-

ration set in, the legs became too weak to support him and he has remained bed-ridden to the present time (13 years after operation).

Case 5.—A man aged 62, an estate manager, three weeks before admission suddenly developed severe pain in the lumbar region radiating down both legs; it was particularly bad in the front of the thighs. He continued to work for five days when numbness appeared in the soles of both feet and spread rapidly up the legs and at the same time the legs became weak and his balance unsteady. There was no sphincter disturbance and no history of spinal injury.

Neurological Examination.—There was considerable weakness of the legs, more marked in the right, so that he was unable to lift them off the bed. One inch wasting of the right thigh and fibrillation in both calves, with general loss of tone, was also seen.

Knee jerks were diminished, ankle jerks absent, and plantar responses extensor.

Sensory change was confined to the antero-lateral aspects of both legs (fourth lumbar dermatome) where pain, temperature, and light touch were diminished. Joint sense was reduced in toes and ankles.

Radiography.—Considerable arthritic lipping of the lower six thoracic vertebrae was shown. There was dense calcification in the nucleus of the eighth-ninth disc with a thin plaque of calcification in the arachnoid, 1 cm. long by 1 mm. thick, at the level of the first lumbar vertebra.

Myelography.—A complete block at D.12/L.1 disc was seen.

Lumbar Puncture.—This showed an absolute manometric block. The fluid was yellow containing 7 cells per c.mm. and 175 mg. of protein per 100 ml.

Operation.—Laminectomy of the twelfth thoracic and first lumbar vertebrae was performed. The spinal cord was pushed over to the right by a large prolapse from the 12/1 disc. The protrusion was exposed extradurally and, surprisingly, in view of the short history, was found to be of an extremely hard consistency and had to be cut away piecemeal. The calcified arachnoidal plaque was also removed.

Post-operative Course.—The patient had total motor and sensory paraplegia below the level of the second lumbar segment. He died four months later in another hospital from pyelonephritis without any recovery taking place. No necropsy was obtained.

Case 6.—A woman aged 44, a housewife, 14 months before admission found that her legs suddenly gave way and on picking herself up she realized that they were weak. The weakness progressed steadily after this, more marked in the right leg, and walking became difficult and she often fell. For six weeks before admission she could only go about by clinging on to furniture for support. At some time during these 14 months numbness had appeared in the legs but she could not give the precise time. She had no sphincter disturbance, no pain at any time, and no history of injury.

Neurological Examination.—There was considerable generalized weakness of both legs, more marked in the right, with increased tone and ankle and patella clonus.

The lower abdominal reflexes were absent, the knee and ankle jerks exaggerated, and plantar responses extensor.

There was a well-defined level at the eighth thoracic dermatome below which there was reduced sensation to pain and temperature, and from the eleventh dermatome downwards there was analgesia and thermoaesthesia. There was only vague and patchy loss to light touch. Joint sense was absent at the toes. Vibration sense was absent up to and including the iliac crests.

Radiography.—Granular calcification in the nucleus of the eighth-ninth disc without narrowing (Fig. 4) was seen. In the antero-posterior view a faint circular rim of calcification, 1·5 cm. in diameter, was visible in the spinal canal overlying the calcified disc (Fig. 5). It could not be seen in the lateral view.

Myelography.—A circular filling defect, 1·5 cm. in diameter, was seen at the calcified disc level and outlining the circular opacity.

Lumbar Puncture.—A complete manometric block was shown. The fluid was clear, containing 2 cells per c.mm. and 95 mg. of protein per 100 ml.

Operation.—Laminectomy o the eighth and ninth thoracic vertebrae was performed. A hard swelling was felt anteriorly in the dural sac. The cord was kinked backwards by a prolapse from the eighth-ninth disc. Owing to the firm consistency of the prolapse it was not removed. Two ligamenta denticulata were divided and the dura left open.

Post-operative Course.—There was total transection below the eighth thoracic segment. At six months a few degrees of movement had returned in the left ankle and knee and in the right ankle with a little recovery of pain sense below the right knee with hyperpathia. The legs were extremely spastic with frequent flexor spasms. The patient had recurring attacks of urinary sepsis and developed bed sores. She eventually died nine months after operation.

At necropsy the prolapse, cord, and dura were removed in one piece and sectioned (Fig. 9).

Case 7.—A woman aged 63, a housewife, 10 years before admission twisted her spine suddenly and the following day she found she was unsteady on her feet tending to veer to the left. A week later on getting into a bath she observed that she could not appreciate temperature in the skin below the knee on the right, and urgency of micturition developed. Two years later both legs became stiff and weak, more marked in the right, but the bladder symptoms had cleared up. During the last six years preceding admission the weakness and stiffness progressed so that she could get about only by holding on to the furniture, and both legs felt numb. For one year the legs were much weaker and she could

just stand with support. Flexor spasms occurred frequently. Urgency of micturition had reappeared with occasional incontinence. There had been no pain at any time.

Neurological Examination.—There was extreme weakness of both legs with a small range of movement retained at all joints, and gross spasticity with ankle and patella clonus.

The abdominal reflexes were absent, the knee and ankle jerks exaggerated, with bilateral extensor plantar responses.

Sensation was difficult to define, but repeated testing indicated some reduction of pain, temperature, and light touch below the second lumbar dermatome on the right and the third dermatome on the left. Vibration sense was absent in the tibiae and patellae. Joint sense was unaffected.

Radiography.—There was gross arthritic lipping of all thoracic vertebrae. No calcification was seen.

Myelography.—Myelograms showed a partial block at the level of the tenth-eleventh intervertebral disc, with a convex lower border.

Lumbar Puncture.—This demonstrated a complete manometric block. The fluid was clear, containing 3 cells per c.mm. and 150 mg. of protein per 100 ml.

Operation.—Laminectomy of the tenth, eleventh, and twelfth vertebrae was performed. A hard lump was palpable. The cord was bulged backwards and was of paper thinness and for almost a centimetre of its length had a translucent blue discoloration. The prolapse was an extremely hard bar extending across the spinal canal containing a small amount of soft cartilage set between bony spurs. The ligamenta denticulata were divided above and below but no attempt was made to remove the disc prolapse.

Post-operative Course.—Paraplegia below the first lumbar spinal segment was complete. It is now four years since operation and there has been no recovery in power or sensation. There is some return of tone in the legs and the patient is able to get about in callipers and crutches. By emptying the bladder every three hours she remains dry.

Case 8.—A man aged 54, a charge hand, 11 weeks before admission suddenly developed severe pain in the lumbar region which radiated down his legs both back and front to the feet. Sharp exacerbations occurred on coughing, sneezing, and straining. He remained in bed for six days and then on getting up found his legs were very weak. This weakness progressed rapidly so that within another few days his legs would not support him. At the same time a burning sensation appeared in the lateral toes of both feet which spread to involve the front of the legs below the knees and within a few days was replaced by numbness. Urgency of micturition appeared at the start of symptoms but cleared up within a few weeks. He was admitted to another hospital five weeks after the onset where after an initial slight improvement his condition remained stationary.

Neurological Examination.—There was considerable generalized loss of power in both legs, more marked in the right. Tone was increased with ankle clonus.

The abdominal reflexes were absent, the knee and ankle jerks exaggerated, and plantar responses extensor.

Cutaneous sensory change was confined to an area on the anterior aspects of both shins (corresponding to the fourth lumbar dermatome) where there was reduction of pain, temperature, and light touch. Position sense was impaired in the toes. Vibration sense was reduced in the left tibia.

He could walk without support but unsteadily, on a broad, shuffling base, with considerable pain.

Radiography.—Gross arthritic lipping of most of the thoracic and all the lumbar vertebrae was seen, but no calcification.

Myelography.—Myelograms showed complete block at the level of the intervertebral disc between D12/L1 vertebrae. The obstruction was outlined from above and below and was found to be 1·5 cm. long and to have a rounded outline.

Lumbar Puncture.—Almost complete manometric block was shown. The fluid contained 2 cells per c.mm. and 200 mg. of protein per 100 ml.

Operation.—Laminectomy of the twelfth thoracic and first lumbar vertebrae was performed. A firm knob was felt anteriorly in the theca. The cord was bulged backwards and was pinker than usual. Anterior to it was a large and extremely hard prolapse from the 12/1 disc. Its hardness was surprising in view of the short history. No attempt was made to remove it and the patient was left with a decompression.

Post-operative Course.—The paraplegia was made worse and retention of urine developed. After 10 days improvement started and within a few weeks he regained his pre-operative level. The cutaneous sensory change cleared up.

Two and a half years after operation he can get about his house and garden and can walk distances up to 200 yards. He can stand for periods up to half an hour before aching in the legs becomes severe. Sphincter control is normal. The operation seems to have arrested the progress of the condition and there has been a little improvement in movement compared with his pre-operative state.

Case 9.—A man aged 61, a labourer, 13 days before admission, while putting on his overcoat, suddenly developed severe pain in the midline of the lumbar region and at the same time his legs became weak. He retired to bed and on attempting to get up two days later he found his legs would not support him. He had observed no sensory or sphincter disturbance, and there was no history of spinal injury.

Neurological Examination.—There was generalized weakness of both legs, more marked in the right, and increased tone with patella clonus. Coordination was grossly impaired.

Both knee jerks were exaggerated, the ankle jerks brisk. Plantar responses were extensor.

There was vague loss of sensation to pin prick below mid-thighs which cleared up within a few days. Temperature and light touch were unaffected. Position sense was considerably reduced at the toes and ankles and vibration sense was absent in the tibiae.

Radiography.—There was arthritic lipping of the lower thoracic and all the lumbar vertebrae, but no calcification or disc narrowing.

Myelography.—A circular lesion causing a complete block was outlined from above and below over the tenth-eleventh intervertebral disc (Fig. 6). In the lateral view a suggestion of a " double profile " could be seen.

Lumbar Puncture.—A partial block was shown. The fluid contained 4 cells per c.mm. and 100 mg. of protein per 100 ml.

Operation.—Laminectomy of the tenth, eleventh, and twelfth thoracic vertebrae was performed. A firm mass could be felt anteriorly in the theca. The cord was kinked backwards and a little swollen but not discoloured. A tense swelling could be seen arising from the 10/11 disc lying to the left of the midline. It was approached extradurally and found to be the size of a pea and composed of soft degenerate disc tissue identical with the common lumbar protrusions. It was removed completely.

Post-operative Course.—After a transient exacerbation of signs power increased considerably in the legs with disappearance of spasticity and clonus and he could walk with support. Joint sense returned to normal within a fortnight. Now, 10 months after operation, he is able to walk any distance with one stick and his only complaint is of a little unsteadiness on his feet. There is no pain or sensory disturbance and sphincter control is normal.

Case 10.—A man aged 44, an engineer, five months before admission developed difficulty in starting micturition and had to strain to empty his bladder. This symptom cleared up in a fortnight. Two weeks later stiffness and weakness of the left leg appeared and after a further month the right leg was similarly affected. The weakness in both legs progressed steadily so that for the month before admission he had been unable to walk and had also become incontinent of urine and faeces. A sensation of coldness behind both knees appeared at the inception of his symptoms and remained stationary until the last few weeks when numbness of the skin below the umbilicus was noticed and which rapidly spread down to involve both legs. For one week there had been frequent flexor spasms. There was no complaint of pain at any time.

Neurological Examination.—Some weakness of the lower abdominal muscles with gross reduction of power in both legs was perceived so that the patient was unable to lift them off the bed, but a small range of movement was retained at all joints. Tone was increased with clonus at the left ankle. The lower abdominal reflexes were absent, the knee and ankle jerks exaggerated, the plantar responses extensor.

There was a level at the eleventh thoracic dermatome on the right and the twelfth on the left below which pain, temperature, and light touch were impaired but not lost. Position sense was absent at the toes, ankles, and knees, and reduced at the hip joints. Vibration sense was lost below the third lumbar spinous process.

Radiography.—Arthritic lipping of the lower three thoracic vertebrae was visible. Calcification of the nucleus pulposus was present in the four intervertebral discs between the 8/9, 9/10, 10/11, and 11/12 vertebrae (Fig. 2).

Myelography.—There was no block discernible on running the contrast up to the cervical region, but in the reverse direction there was a transient arrest at the 6/7 disc and again at the 8/9.

Lumbar Puncture.—Partial block was shown. The cerebrospinal fluid was clear and contained 2 cells per c.mm. and 20 mg. of protein per 100 ml.

Operation.—Laminectomy of the seventh to the tenth vertebrae inclusive was performed to expose both levels which the myelogram had shown to be abnormal. The protrusion was found at the lower, 9/10 level (there was no abnormality discovered at the 6/7 disc). The cord was paler than normal and had fewer blood vessels on its surface. It was kinked back by a protrusion the size of a pea lying slightly to the left of the midline. The centre of the prolapse was soft but its wall was very hard and calcified, and it appeared that the protrusion had been present for a much longer time than the symptoms suggested. Its contents were removed and then the rim excised piecemeal with a small scoop.

Post-operative Course.—Transient worsening of physical signs for a few days was followed by steady if slow improvement. Within two weeks the patient could lift his legs off the bed and had regained bladder control. The sensory and reflex changes persisted. Two and a half years after operation he can walk up to 500 yards with two sticks and can climb stairs slowly. He has recently returned to sedentary work. He still has some urgency of micturition.

Case 11.—A man aged 45, a research chemist, whose clinical features have been described on a previous page.

Radiology.—Calcification was present in the nucleus pulposus of the ninth-tenth disc, without any narrowing (Fig. 3).

Myelography.—Myelograms showed an oval filling defect at the level of the calcification measuring 1½cm. in diameter (Fig. 7).

Lumbar Puncture.—Manometrics were normal. The cerebrospinal fluid contained 4 cells per c.mm. and 50 mg. of protein per 100 ml.

Operation.—In view of the discrepancy between the clinical and radiological levels it was thought there might be two lesions and that both should be explored.

The upper level was exposed first and revealed no abnormality. A laminectomy of the ninth and tenth vertebrae was performed and a firm swelling could be felt in the dural sac On opening the theca the cord was seen to be displaced backwards by a prolapsed disc. The protrusion arose by a narrow neck and then expanded into a spherical mass about 1½ cm. across with the dura and cord stretched over its surface. There was no discoloration of the cord substance. The rim of the disc was hard and partly calcified but its contents were soft, degenerate cartilage. As a result of the intimate relationship to the cord it was extremely difficult to remove all the calcified rim and its right-hand margin had to be left *in situ*.

Post-operative Course.—Physical signs were made considerably worse and the bladder required tidal drainage. Improvement started after 10 days. By the time of discharge from hospital five weeks later power had improved so that the patient could walk without assistance although unsteadily. By four months after operation he was walking normally.

Three and a half years after operation he walks, runs, swims, and drives a car. All modalities of sensation are normal except for subjective numbness of the feet. The right knee jerk is still brisk but plantar responses are flexor. His only complaint is that of a sensation of tightness in the calf muscles.

Summary

Attention is drawn to the serious damage inflicted on the spinal cord by central thoracic disc protrusions. Eleven cases are described and it is shown that this type of prolapse is confined to the lower nine disc spaces without any particular level predominating. It is commoner in men of middle and late adult life and has little relationship to trauma.

The clinical features are not characteristic and are those of a fairly rapidly progressive spinal compression, so that in eight of the patients a severe neurological disability leading to operation was present within seven months from the onset of symptoms. Three cases presented with incomplete Brown-Séquard syndromes. Pain was an infrequent symptom. Sphincter disturbance occurred late and was present in less than half the cases.

Five patients showed a manometric spinal block with high protein but the important feature was the number of patients with only a slight rise of protein or little evidence of block. *X*-ray changes formed the important diagnostic guide. Eight showed calcification of the nucleus pulposus. The typical myelographic appearance was an oval filling defect.

The dangers associated with visible degenerative changes in the cord are emphasized.

The pathological basis of interference with cord function produced by the small protrusions is discussed.

Operation results are reviewed. Three patients developed total transections post-operatively and two of them died. Two other patients were made worse by surgery and made slow, incomplete recoveries. Six patients did well, five of them returned to work, and in three the improvement was of a grade similar to that following removal of benign spinal neoplasms such as meningiomata or neurofibromata. Some details of surgical technique are stressed.

It is a pleasure to express my indebtedness to my senior colleague Mr. Wylie McKissock for permission to include six of his patients in this series ; to Professor T. Crawford for the preparation of the necropsy specimen in Case 6 and the photomicrographs of the sections, and to Dr. James Bull for many of the radiographs and myelograms.

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STUDIES OF ISCHAEMIC AND POST-ISCHAEMIC PARAESTHESIAE IN NORMAL SUBJECTS AND IN SCIATICA

BY

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The occurrence of paraesthesiae during occlusion of the circulation to a limb (ischaemic paraesthesiae) and after its restoration (post-ischaemic paraesthesiae) is well recognized. The ischaemic paraesthesiae consist of a soft, diffuse, tingling sensation felt at the periphery of the limb and lasting only a few minutes. The post-ischaemic paraesthesiae, which have been well demarcated by Merrington and Nathan (1949), are of four types : thermal, which are hot and cold sensations felt soon after restoration of the circulation to the limb ; tingling, a soft, diffuse, vibrating feeling ; pricking, composed of numerous, irregular, fine, discrete pricks ; and pseudo-cramp, in which a sensation of movement in and tension of the muscle is experienced.

The site of origin of these phenomena has been the subject of differing opinions. Weddell and Sinclair (1947) thought that, though the ischaemic paraesthesiae arose from the nerve trunk, the post-ischaemic paraesthesiae had their origin in the sensory end-organs. Lewis, Pickering, and Rothschild (1931), Bazett and McGlone (1932), and Zotterman (1933), on the other hand, have all considered that the post-ischaemic paraesthesiae arise in the nerve trunk. The careful studies of Merrington and Nathan (1949) have indicated that this latter view is correct.

The primary purpose of the present work was to study the development of ischaemic and post-ischaemic paraesthesiae in the legs of patients suffering from sciatic pain due to a prolapsed intervertebral disc. Previous studies of ischaemic and post-ischaemic paraesthesiae have been concerned with the upper limb. It was, therefore, necessary first to study the development of these phenomena in the lower limbs of normal subjects to establish norms.

Material and Methods

The studies on subjects with sciatica were made on 21 patients with unilateral sciatic pain believed to be due to prolapse of an intervertebral disc. Six of these patients came to operation and a prolapsed disc was found. The studies on normal subjects were made on a series of volunteers, some accustomed to clinical experiments, others entirely unused to such procedures. All the observations were made in the following way. The subject lay supine and a sphygmomanometer cuff 13 cm. wide was placed around the thigh with its lower margin 7·5 cm. above the upper border of the patella. The limb was elevated to empty the veins and then the pressure in the cuff was rapidly raised to 200 mm. of mercury. The limb was then lowered to the horizontal and the occlusion maintained for 20 minutes. The time of onset of the ischaemic paraesthesiae was measured from the moment of occluding the circulation. The time of onset of the various post-ischaemic paraesthesiae was measured from the moment of the release of the occlusion. All tests were done at room temperature, and no limb was retested within 24 hours of a previous test. When both legs of a subject were tested they were done separately so as to facilitate unbiased observation of each limb.

The classification of the paraesthesiae used here is that of Merrington and Nathan described above. Attention was concentrated on the ischaemic paraesthesiae and on the thermal, tingling, and pricking types of post-ischaemic paraesthesiae. With a period of occlusion of 20 minutes pseudo-cramp occurs inconstantly and so was not studied. Because of the subjective difficulty of recognizing the disappearance of the paraesthesiae or of assessing their severity, interest was mainly devoted to the time of onset of the phenomena and not to their duration or intensity.

Results

Occlusion of the circulation to the lower limb for 20 minutes and its subsequent restoration produced exactly the same kinds of ischaemic and post-ischaemic paraesthesiae as have been described by Merrington and Nathan for the upper limb. This similarity covered the nature of the sensations, their

time and order of onset, duration, and their modifiability by deformation of the skin and by pressure. The first point of interest was, however, that all forms of paraesthesiae were not felt on every occasion by all subjects. The frequency with which each type was experienced in a series of 97 tests is shown in Table I. Such omissions do not appear to

TABLE I

INCIDENCE OF THE VARIOUS PARAESTHESIAE IN 97 TESTS

Ischaemic	79
Thermal	88
Tingling	93
Pricking	78

have been found in the arm except by modification of the experimental conditions. Some of the omissions might be explained by a failure of observation on the part of the subject, but this cannot be the whole explanation, for in several instances paraesthesiae of one type not felt on one occasion were experienced on another. Thus seven subjects who observed tingling but not pricking at one test experienced both paraesthesiae at another, and this not always a subsequent occasion. One subject especially felt only thermal paraesthesiae in both legs. It seems, therefore, that the occurrence of all types of paraesthesiae in every subject on all occasions is not invariable. Variations occur from time to time even in the same subject under identical experimental conditions.

To assess the constancy of the time of onset of the various paraesthesiae in a large series of tests in normal subjects the means, ranges, and standard deviations were calculated and are given in Table II. It demonstrates clearly that the time of onset of each

TABLE II

TIME OF ONSET OF PARAESTHESIAE AFTER OCCLUSION AND AFTER RESTORATION OF CIRCULATION

Type of Paraesthesia	No. of Tests	Mean (secs.)	Range (secs.)	Standard Deviation
Ischaemic	58	389	60–1140	309
Thermal	65	12·6	3–45	6·3
Tingling	69	46	10–120	6·4
Pricking	75	64	40–270	48·6

form of paraesthesia does not follow a normal distribution curve but shows a wide variation and is not so constant as might be expected from the reports of studies on the upper limb. Repeated tests on the same leg in a series of normal subjects (Table III) did not show any greater constancy in the time of onset of the various paraesthesiae. Despite this wide variation, however, the order of onset of the post-ischaemic paraesthesiae was invariably thermal, tingling, and pricking, and there was not a single

TABLE III

TIME OF ONSET OF PARAESTHESIAE AFTER OCCLUSION OR RESTORATION OF CIRCULATION IN REPEATED TESTS ON A SERIES OF NORMAL SUBJECTS

Time of Onset in First Test (secs.)				Time of Onset in Second Test (secs.)			
Ischaemic	Thermal	Tingling	Pricking	Ischaemic	Thermal	Tingling	Pricking
225	10	20	44	195	5	23	—
810	7	40	120	390	10	—	135
540	—	30	—	622	—	33	45
225	15	35	35	581	16	120	—
150	25	54	70	200	18	40	66
—	45	65	—	60	28	62	100
—	43	65	—	—	14	30	100

exception to this rule. It would seem, therefore, that despite the marked fluctuations in the times of onset of the paraesthesiae there is something fundamental in their order of onset. It was suggested that the variation on repeated testing might be less in a subject accustomed to making experimental observations than in a series of less experienced persons. Table IV shows the results of repeated tests in such a subject. Comparing the results in

TABLE IV

TIME OF ONSET OF PARAESTHESIAE IN A SERIES OF TESTS IN AN EXPERIENCED SUBJECT

Time of Onset in Right Leg (secs.)				Time of Onset in Left Leg (secs.)			
Ischaemic	Thermal	Tingling	Pricking	Ischaemic	Thermal	Tingling	Pricking
180	—	65	80	150	5	45	75
735	15	27	45	—	3	50	70
100	3	47	90	540	7	47	85
120	3	43	85	120	4	43	90
150	5	50	80	100	8	20	45
180	7	50	83	—	9	52	90
120	4	58	87	105	10	44	80
115	7	55	100	—	6	55	85
75	5	47	90	110	9	45	110

Table IV with those in Table III by means of the " F " test, it is found that F is 1·718 for the ischaemic paraesthesiae which is not significant ; F is 47·445 for the thermal paraesthesiae which is significant at the 1% level ; F is 1·159 for tingling which is not significant ; and F is 27·593 for pricking which again is not significant, though in view of the small number of observations, too much stress should not be laid on this. The trained observer, therefore, did not appear to achieve in general a greater degree of test re-test consistency than the ordinary subject.

With these facts in mind a study was made of ischaemic and post-ischaemic paraesthesiae in patients who, at the time of testing, were suffering from sciatic pain due to a prolapsed intervertebral disc. A series of 21 patients with unilateral sciatic pain was tested. The history and examination of each patient was such as to indicate that the sciatica

was due to prolapse of an intervertebral disc. Six of the patients had a laminectomy at which a prolapsed disc was in fact found. The results of the tests are given in Table V. In each case the affected leg was compared with the non-affected leg as control. A statistical analysis of the results by means of Fisher's t test (Table V) showed a significant difference between affected and non-affected legs with regard to the time of onset of the ischaemic

TABLE V

TIME OF ONSET OF PARAESTHESIAE IN A SERIES OF PATIENTS WITH SCIATICA

No.	Time of Onset in Affected Leg (secs.)				Time of Onset in Unaffected Leg (secs.)			
	Ischaemic	Thermal	Tingling	Pricking	Ischaemic	Thermal	Tingling	Pricking
1	65	10	12	60	240	—	40	90
2	300	—	70	—	720	7	40	80
3	180	5	55	70	60	6	30	55
4	45	10	25	70	600	10	10	60
5	175	15	50	80	205	9	45	—
6	125	10	40	55	1140	5	20	40
7	140	10	60	90	180	15	65	70
8	840	12	20	60	720	20	30	210
9	480	8	70	—	230	5	55	—
10	35	10	30	40	110	15	30	60
11	180	15	45	90	225	9	65	270
12	180	10	10	70	1020	14	40	65
13	—	9	45	103	1020	14	59	95
14	195	5	60	60	720	40	75	80
15	165	5	10	—	105	15	40	—
16	105	5	30	195	—	20	75	160
17	80	9	22	49	225	10	20	44
18	900	11	85	110	810	7	40	120
19	300	22	39	50	540	—	30	—
20	540	8	95	160	—	9	110	—
21	180	10	30	62	225	15	35	65

"t" = 2·390 for ischaemic (significant at the 2·5% level), 1·679 for thermal, 0·493 for tingling, and 1·268 for pricking, all of which are not significant.

paraesthesiae, the onset being accelerated on the affected side. No difference was found with regard to the post-ischaemic paraesthesiae. As a further control a series of 12 normal subjects was tested (Table VI) and again compared by the t test but no significant difference was found in either the ischaemic or post-ischaemic paraesthesiae in the two legs.

Examination of the figures for the times of onset of the ischaemic paraesthesiae in the subjects with sciatica (Table V) shows that they can be divided into two groups. In eight cases the length of time before ischaemic paraesthesiae developed in the affected leg was less than half the value for the non-affected leg, whereas in the other 10 cases the time was approximately equal on the two sides. It seems, therefore, that the significant difference between the values on the affected and non-affected sides in the whole group is due to the large difference in these eight cases.

A study of the clinical records in these eight cases showed that in every case there was an indisputable

TABLE VI

TIME OF ONSET OF PARAESTHESIAE IN A SERIES OF NORMAL CONTROLS

Time of Onset in Right Leg (secs.)				Time of Onset in Left Leg (secs.)			
Ischaemic	Thermal	Tingling	Pricking	Ischaemic	Thermal	Tingling	Pricking
660	10	33	—	155	5	10	55
960	14	30	50	300	6	20	70
—	18	—	—	—	25	—	—
—	29	60	—	—	20	62	—
840	15	35	55	420	—	44	52
300	—	46	65	480	20	53	67
910	5	85	110	—	8	75	135
870	20	40	60	900	—	37	47
690	12	31	50	140	15	32	43
180	—	65	80	150	5	45	75
—	14	23	—	—	11	—	—
180	5	65	85	240	5	10	58

"t" = 2·035 for ischaemic, 0·743 for thermal, 1·695 for tingling, and 0·167 for pricking, none of which are significant.

neurological deficit such as loss or diminution of the ankle jerk on the affected side or sensory impairment. Of the other 10 cases, only three had definite neurological signs, the remaining seven cases having sciatic pain and the orthopaedic signs associated with a prolapsed disc. The marked acceleration in the time of onset of the ischaemic paraesthesiae in sciatica due to a prolapsed disc does, therefore, appear to be related to the degree of damage to the nerve as measured by the neurological signs.

Discussion

Though prolapse of an intervertebral disc is well established as a cause of sciatic pain, there is considerable evidence to show that its mode of action is far from simple. Falconer, McGeorge, and Begg (1948) have shown both from myelographic and operative studies that sciatic pain may disappear though the prolapse persists. They also found evidence of delay in the passage of nerve impulses through the affected nerve root. They commented: "The production of sciatica thus depends on the association of two factors, the presence of a disc prolapse and changes within the nerve root." Malcolm (1951) studied the reflex time of reflexes mediated through affected roots and also found a significant delay.

The present studies adduce evidence of there being changes not only in the root but also in the nerve well peripheral to the site of disc compression. Though one might expect delay in the time of onset of post-ischaemic paraesthesiae due to slowing of conduction through the nerve, this method is much too crude a measure to detect any evidence of such a delay. Hence restoration of the blood supply to the affected and non-affected legs reveals no significant difference in the time of onset of the post-ischaemic paraesthesiae. In the case of the ischaemic

paraesthesiae also there is no delay, but in fact their time of onset is accelerated to a significant degree in the affected leg. As the portion of the nerve from which the paraesthesiae arise is well peripheral to the site of disc compression, there is thus evidence of changes in the periphery of the nerve of such a nature as to impair its ability to withstand ischaemia. These studies provide no evidence as to the nature of these changes. Such pathological observations as have been made (Denny-Brown, 1933) have only shown oedema of the nerve trunk, but there may well be other changes in the myelin sheath which impair the functioning of the nerve.

It seems clear, however, that these changes, whatever their nature, are such as to render the nerve more sensitive to one form of insult, namely, ischaemia. This raises the possibility that these changes in the periphery of the nerve may make an important contribution to the symptomatology of the prolapsed disc, especially in the intractable cases which fail to gain relief even from operation. These considerations seem worthy of further study.

Summary

The times of onset of ischaemic and post-ischaemic paraesthesiae have been studied in the lower limb.

Evidence is presented that the onset of ischaemic paraesthesiae is accelerated in a limb in which sciatic pain due to a prolapsed intervertebral disc is present.

Observations are described which suggest that in the presence of a prolapsed intervertebral disc there are important changes in the periphery of the nerve.

I wish to thank Dr. W. Ritchie Russell, Dr. C. W. M. Whitty, and Dr. P. W. Nathan for their advice on this work, and Mr. J. E. A. O'Connell for his kindness in allowing me to examine his patients and for his operative findings. I also wish to thank Brigadier W. R. D. Hamilton, O.B.E., consultant physician, the War Office, for permission to report the findings in the Service patients, and Dr. A. S. C. Ehrenberg and Dr. S. Crown for advice about the statistics.

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THREE CASES OF ACHONDROPLASIA WITH NEUROLOGICAL COMPLICATIONS

BY

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The role played by congenital malformations of the skull and spine in the production of lesions of the central nervous system has been increasingly appreciated in recent years. In the skull premature synostosis of the sutures of the vault (craniostenosis) can lead to optic atrophy and symptoms of intracranial hypertension. Hydrocephalus and compression of the brain stem and upper cervical cord can result from deformities of the base of the skull, such as platybasia and narrowing of the foramen magnum. In the spine congenital deformities, such as hemivertebrae, absence of cervical segments, scoliosis, and kyphosis, may also result in cord compression in adult life.

In this paper attention is drawn to the possible effects on the nervous system of achondroplasia. This condition of foetal chondrodystrophy is well known and is characterized by failure of endochondral ossification. It results in a striking disproportion of the body ; a large head, short limbs, and a spine of relatively normal length.

The complications of hydrocephalus and compression of the spinal cord and cauda equina are demonstrated in the following case reports.

Case 1.—This was a man aged 40, an achondroplasic dwarf. He was 4 ft. 7½ in. high, with a head circumference of 23 in. He was of average intelligence. His father was similarly afflicted and in his forties developed paralysis of the legs, for which a spinal operation was performed in 1913. The patient stated that some recovery took place, but he died a few years later when paralysis of the lower limbs was then complete.

The patient was well until January, 1951, when he began to suffer from pains in the lumbo-sacral region which later radiated down the outer aspects of both thighs. Six months afterwards he felt tingling sensations in both feet and two weeks before admission to hospital in July, 1951, he felt that his buttocks and the soles of his feet were numb. There had been some difficulty in micturition for seven days.

On examination speech was normal. His gait was waddling with noticeable weakness of dorsiflexion of the feet. The cranial nerves were normal. The arms were normal. Abdominal reflexes were present. Power in both legs was considerably reduced, especially below the knees, with bilateral partial foot drop. Knee and ankle jerks were absent on each side. Plantar reflexes were flexor. There was no wasting or fibrillation. Sensation was moderately impaired bilaterally (L.5, S.1, and 2), and analgesia was marked (S.3, 4, and 5) bilaterally. There was lumbar kyphosis (Fig. 1) with slight generalized restriction of spinal movements.

FIG. 1.—Gibbus deformity of lumbar spine in Case 1.

Lumbar puncture at L.4–5 showed clear orange coloured fluid (protein 2,060 mg. per 100 ml.) and absolute block on bilateral jugular compression.

Radiological Examination.—The patient had a fairly large head, with a shallow posterior fossa, a normal foramen magnum, no platybasia, and small facial bones with a slight degree of hypertelorism.

Cisternal air encephalography showed slight sym-

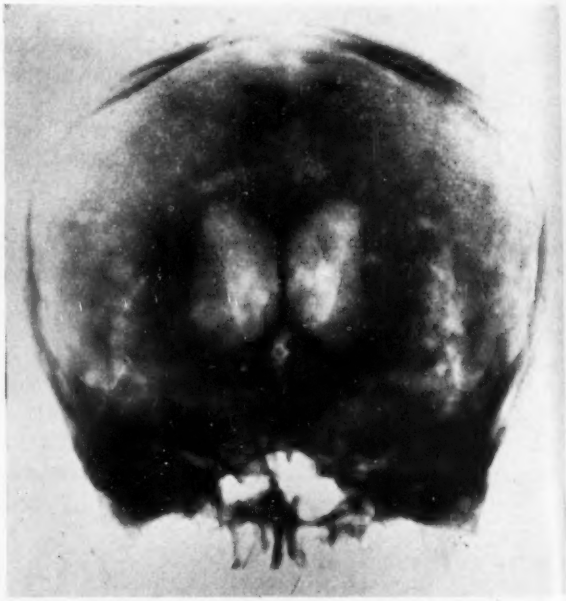

FIG. 2.—Air encephalogram showing dilatation of lateral ventricles in Case 1.

metrical ventricular dilatation (Fig. 2) including the third ventricle. The aqueduct and fourth ventricles were normal.

The cervical spine was normal, except for loss of the lordotic curve. The dorsal spine was normal. The lumbar spine had marked kyphosis centred at L.2 (Fig. 3), the body of which vertebra was slightly wedged in the lateral view. The posterior surfaces of the lumbar vertebrae were hollowed out, producing an undulating anterior margin of the spinal canal. The intervertebral disc spaces were abnormally wide. There was no spondylolisthesis. The interpedicular widths were normal and it was not possible to demonstrate any shortening of the pedicles.

FIG. 3.—Lumbar spine; kyphosis, wedging of L.V.2, wide disc spaces, and concave posterior borders of vertebral bodies in Case 1.

Lumbar Myelography.—The myodil collected in three pools opposite the bodies of L.3, 4, and 5 (Figs. 4a and b). Tilting to 80° demonstrated that these pools of myodil were fixed. Subsequent examination confirmed the evidence of block at these levels. It was concluded that there was cauda equina compression opposite the bodies and discs L.3, 4, and 5.

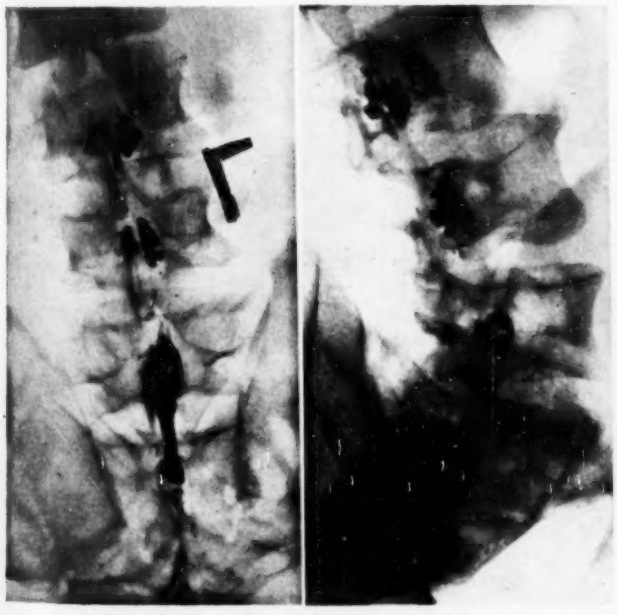

FIG. 4a and b.—Lumbar myelogram; myodil arrested opposite bodies and discs of L.V. 3, 4, and 5 in Case 1.

Laminectomy.—Professor Lambert Rogers performed a laminectomy at L.2, 3, 4, and 5. The dural tube was displaced posteriorly at L.2, 3, and 4 by massively protruding intervertebral discs which were removed. The dura was then opened and the cauda equina found to be slack and relieved of pressure.

Progress.—Following this operation the patient made good progress and has now returned to his work and regained good function in both legs. The reflexes are still absent but the sensory disturbance has practically cleared.

Case 2.—This woman was aged 37 years and was a typical achondroplasic dwarf (Fig. 5), standing 4 ft. 5 in. high with a head circumference of 24 in. She was of good intelligence, and had no family history of dwarfism.

She was well until nine years ago when she first noticed numbness of the left foot. Subsequently she developed low back pain with radiation of sciatic distribution. Weakness of the legs developed two years ago with bilateral foot drop. She had no symptoms referable to the head, trunk, neck, or upper limbs. Precipitancy and frequency of micturition were present. She had worn a caliper on the left leg for the past year.

FIG. 5.—Slight dorsolumbar kyphosis and horizontal sacrum in Case 2.

On examination it was seen that she was only able to walk slowly with the aid of two sticks. Speech was normal. The cranial nerves were normal. The upper limbs were normal. The upper abdominal reflex was present, the lower reduced. There was bilateral foot drop with corresponding muscular atrophy and loss of both ankle jerks. The right knee jerk was reduced, the left normal. Plantar reflexes were flexor. All movements at the ankle were weak, but those at the hip and knee were nearly normal. All forms of cutaneous sensation below both knees were markedly impaired. Between the knees and groin pin-prick and temperature differences were not fully appreciated. Vibration and joint position sense were grossly defective in the feet.

Lumbar puncture at L.3–4 demonstrated absolute manometric block on bilateral jugular compression. The cerebrospinal protein was 575 mg. per 100 ml.

Radiological Examination.—The skull was large, with a normal foramen magnum and no platybasia.

Cisternal air encephalography showed marked symmetrical dilatation of the lateral ventricles (Fig. 6) with

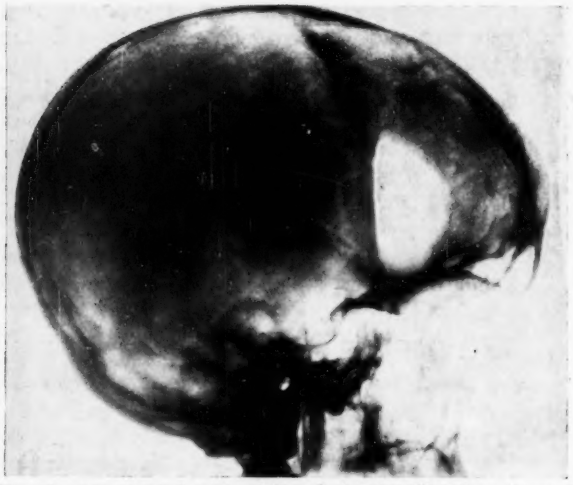

FIG. 6.—Air encephalogram of Case 2 showing dilatation of lateral ventricles.

some dilatation also of the third ventricle, the aqueduct, and the fourth ventricle. No other deformity or displacement was shown. In the cervical spine the vertebrae were tall but there was no other abnormality. There was lower dorsal lordosis only. There was slight dorso-lumbar kyphosis (Fig. 7) with exaggerated lower lumbar lordosis. The posterior surfaces of the vertebral bodies were concave with large intervertebral disc spaces, and osteophytes at L.1–2 and L. 2–3. The sacrum lay horizontally. Pedicles and interpedicular widths were normal.

FIG. 7.—Lumbar myelogram showing slight dorso-lumbar kyphosis, lumbar lordosis, concave posterior borders of vertebral bodies, wide disc spaces anteriorly, myodil arrested at L.V.4 with complete block at L.V.3 in Case 2.

Lumbar myelography showed partial block at L.4 and complete block at L.3.

Myodil was arrested at the L.1–2 disc space. A pool of myodil collected opposite each vertebral body from D.8 to L.1, and the myodil column was interrupted opposite each disc between these levels (Figs. 8a and b).

It was concluded that there were multiple intervertebral disc protrusions with cauda equina compression.

Laminectomy.—A laminectomy performed by Mr. Charles Langmaid disclosed a narrow spinal canal with multiple disc protrusions and cauda equina compression. The protruding material was removed.

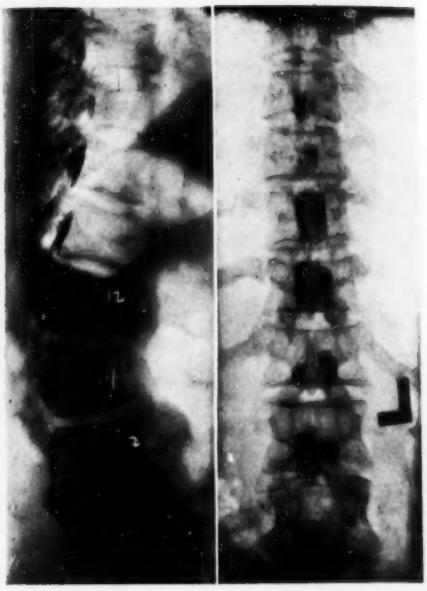

FIG. 8a and b.—Cisternal myelogram of Case 2 showing myodil arrested at L.V.1-2 disc space with pool of myodil opposite each vertebral body from D.8 to L.1.

The patient's subsequent progress was slow. She has not regained power in the left foot but there is moderate improvement in the power of the right lower limb and her gait is better.

Case 3.—This was a man aged 29, a typical achondroplasic dwarf, standing 4 ft. 10 in. high, with a head circumference of 24·5 in. His intelligence was below average. He had no family history of dwarfism.

The patient was well until two years ago when he first began to notice weakness and stiffness of the lower limbs. At first there was dragging and periodic trembling of the left leg. Later both legs became weak and he found increasing difficulty in walking. He was forced to give up his work three months ago. Recently he had had some pain in the cervico-occipital region and periodic substernal discomfort.

On examination it was seen that he was unable to walk without assistance and had a spastic tetraplegia. The cranial nerves were normal. Neck movements were normal. There was slight upper lumbar kyphosis. Power in both arms was markedly reduced but in the legs power was little affected. The biceps, triceps, and radial reflexes were grossly exaggerated, with brisk finger reflexes. Abdominal reflexes were absent and there was bilateral knee and ankle clonus. There were extensor plantar responses bilaterally. There was no obvious wasting. Coordination was normal. Cutaneous sensation was considerably reduced from C.2 to D.1, but below this level there was only slight loss of superficial sensibility. Vibration sensation was absent in the lower limbs and not normally perceived below mid-thoracic level. It was also reduced in the fingers and hands.

Lumbar puncture at L.4–5 gave a clear fluid with normal manometric tests when the head was in the mid-position. When the head was either flexed or extended more than 20° from the midline position complete spinal manometric block was revealed. Cerebrospinal protein was 280 mg. per 100 ml.

Radiological Examination.—The skull had a large vault, the base smaller than normal (Fig. 9). The clivus was steep and short. The posterior fossa was shallow. The squamous occiput appeared to overhang the atlas. There was slight hypertelorism, but no evidence of platybasia. The foramen magnum was normal.

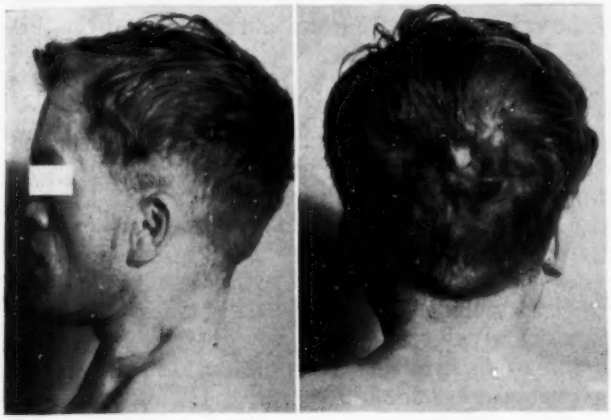

FIG. 9.—Large head, depressed nasal bridge, short neck of Case 3.

A lumbar air encephalogram showed moderate symmetrical dilatation of the lateral ventricles (Fig. 10) and slight dilatation of the aqueduct and third ventricle. Filling of the fourth ventricle in the brow-down position was not possible.

The vertebral bodies were shallow and broad (resembling an infantile type of spine). With flexion of the head there was subluxation of the axis on C.3. The posterior surface of the body of the axis was 4 mm. anterior to that of C.3. There was slight dorsal lordosis and upper lumbar kyphosis (Fig. 11). The L.2 vertebra was wedge-shaped. The pedicles and interpedicular widths were normal. The posterior surfaces of the bodies of the lumbar vertebrae were concave and the intervertebral disc spaces were large.

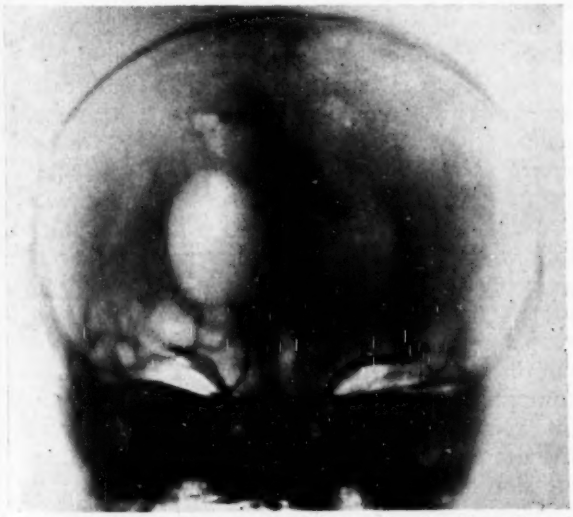

FIG. 10.—Air encephalogram of Case 3 showing dilatation of lateral ventricles.

FIG. 11.—Lumbar spine showing upper lumbar kyphosis, wedging of L.V.2, concave posterior borders of vertebral bodies which are shallow and broad in Case 3.

Lumbar Myelography.—In the lumbar region there was constriction of the myodil column opposite each intervertebral disc space but without any actual hold-up. In the cervical region there was indentation of the anterior surface of the myodil column opposite each disc space. Confirmation of the presence of subluxation of the axis on C.3 was obtained (Fig. 12). The myodil outlining the anterior wall of the thecal space was separated from the posterior surface of the axis by a distance of 5 mm., as compared with 2 mm. between it and the posterior surface of the body of C.3. No bony exostosis could be demonstrated.

Discussion

Paraplegia.—There has been some dispute about the condition of the vertebral column in achondroplasia. Some authors have described only the characteristic lumbar lordosis and tilting of the sacrum but Breus and Kolisko (1900) and Donath and Vogl (1925) showed that abnormalities of the spine were a regular occurrence and that there were changes in the individual vertebrae and in the spine as a whole. They noted a decrease in the height of the vertebral bodies and shortness of the pedicles. The latter resulted from premature synostosis of the centres of ossification of the body with those of the lamina. The spinal canal was narrowed in one or all of its diameters and this led to tight encasement of the normally developed spinal cord. Donath and Vogl also pointed out that in achondro-

FIG. 12.—Lumbar myelogram of Case 3 showing subluxation of atlas on C.V.3.

plasia the normal developmental " unrolling " of the spine is incomplete owing to the absence of a well organized and directed growth of cartilage ; straightening is not completed. The infant with achondroplasia is born with some degree of dorso-lumbar kyphosis and a gibbus may develop during life. They reported the case of a 57-year-old male adult achondroplasic who developed a spastic paraplegia with urinary retention. The first lumbar vertebra was wedge-shaped and displaced posteriorly so that the twelfth thoracic and second lumbar vertebrae articulated with each other anteriorly. Spinal manometry at L. 4–5 was normal ; the cerebrospinal fluid was normal. The patient died four years later. At necropsy the constriction of the spinal canal was pronounced and resulted from shortness of the pedicles and " mushrooming " of the superior articular process and encroachment on the spinal canal. The cord itself was indented and softened *above* the gibbus.

In 1949 Vogl and Osborne reported another case of cord compression in an achondroplasic male aged 30 years. He developed pain in the back and legs, unsteadiness of gait, urinary incontinence, and sexual impotence. There were no definite motor or sensory changes but the knee and ankle jerks were hyperactive. The twelfth thoracic vertebra was wedged and there was marked osteophyte formation between the eleventh and twelfth vertebrae. Lumbar puncture (L.2–3) showed that there was partial manometric block ; the cerebrospinal fluid was 80 mg. per 100 ml. Myeloscopy disclosed narrowing of the spinal canal. Opaque myelography indicated partial block at Th.11–12. Laminectomy confirmed these findings and was followed by relief of symptoms.

Up to 1949 Vogl and Osborne said that there had been 10 published cases (including their own) of " transverse myelopathy " in achondroplasic dwarfs. Few of these reports are at all complete. I have come across one further report by Poynton (1907) in Allbutt and Rolleston's *System of Medicine*. Poynton's patient was 15 years of age, 41 in. in height, and began to develop a spastic paraplegia in his thirteenth year. His mental state was good. The cause of the paraplegia cannot be deduced in the case reports of Parhon and Schunda (1913) and of Parhon and Zalpachta (1905), two cases ; of Zosin (1910), of Falta (1913), of Maas (1920), and of Keményffi (1924).

In the two remaining reports neurological and radiological findings are recorded ; manometry and myelography were not performed.

Albrecht and Ranzi (1926) published the case of an achondroplasic dwarf, aged 46 years, who developed complete spastic paraplegia with incontinence over a 10-year period. Lumbar puncture yielded repeated dry taps and there was wedging with osteophyte formation of the twelfth thoracic vertebra. Laminectomy (Th.10, 11, and 12) confirmed the presence of cord compression ; the spinal canal appeared too tight for the dural sac which did not pulsate and bulged from the operative gap. There was considerable improvement following this decompression laminectomy.

In the second case (Freund, 1933) the patient, aged 21 years, developed spastic weakness of the lower limbs, with pain in the back and thighs, over a period of one year. There was sensory loss below L.3 on the left side. Sphincters were unaffected. Lumbar puncture was not done. The second lumbar vertebra was wedge-shaped and there was a severe gibbus deformity. Traction gave temporary relief. Laminectomy (Th.12–L2) revealed severe angulation of the cord and cauda equina, with a non-pulsatile tight dura. The latter was not opened. A flaccid paraplegia with incontinence followed the operation and a second exploration was undertaken to exclude the presence of a haematoma. None was found. The author felt that a simple congenital stenosis of the spinal canal was not the explanation for the paraplegia, but that cord compression was subsequent to encroachment of the spinal canal by marginal exostoses. As in Donath and Vogl's case, the site of the cord compression was *above* the level of the gibbus.

Neurological complications may also arise in the condition known as hereditary deforming

dyschondroplasia (Slepian and Hamby, 1951). Compression of the spinal cord, the brachial plexus, and various nerves by bony exostoses have been recorded.

In Cases 1 and 2 of the present report cauda equina compression occurred at several levels as a result of massive protrusion of lumbar intervertebral discs. In Case 3 there was subluxation of the second cervical vertebra upon the third, and manometric spinal block was produced by slight flexion or extension of the neck. In the first two cases the bodies of the cervical vertebrae were rather tall. In all three the posterior surfaces of the bodies of the lumbar vertebrae were concave and the adjacent intervertebral disc spaces were abnormally wide. These appearances in the cervical and lumbar spine were also observed in five out of six further achondroplasic adults. One of these complained of pain and stiffness in the back and thighs but there were no neurological abnormalities. In another case the right knee and ankle jerks were markedly exaggerated and the plantar reflex on that side was questionably extensor. In a third case there was an isolated extensor plantar response but it was not constantly demonstrable.

Hydrocephalus.—Marie (1900) considered that the enlargement of the head was probably due to the presence of hydrocephalus although he lacked necropsy evidence. Jansen (1912) concluded that shortening of the base of the skull involved the oral cavity and the sagittal dimensions of the foramen magnum. He quoted Virchow, who considered that kyphosis basis cranii was a characteristic feature of the disease. Jansen showed that the brachycephalic configuration of the skull in achondroplasia was also contributed by the increased width of the skull. Two of his five cases had hydrocephalus which he attributed to an impaired outflow of cerebrospinal fluid from the ventricular system caused by brain stem compression and distortion. Dandy (1921) supported this explanation. Hydrocephalus was present in his two cases and in one was demonstrated by ventriculography to be of the communicating type. He thought that the chondrodystrophy at the base of the skull causes obstruction in the posterior fossa and the development of ventricular dilatation. In some cases it has been demonstrated that the hydrocephalus is present at birth (Russell, 1949), and Dandy suggested that growth of the base of the skull after birth might lead to some relief of the hydrocephalus. In a recent examination of an achondroplasic infant, stillborn at the eighth month, I was unable to find any evidence of hydrocephalus.

In each of the three cases which form the basis of the present report, air encephalography revealed the presence of internal hydrocephalus. The dilatation of the lateral ventricles was symmetrical and was slight in Case 1, marked in Case 2, and moderate in Case 3. In Case 1 the dilatation involved the third ventricle; the aqueduct and the fourth ventricle were normal. In Case 2 the third and fourth ventricles and the aqueduct were involved in the dilatation. In Case 3 the aqueduct and third ventricle were dilated but filling of the fourth ventricle could not be obtained. Intelligence was average in Case 1, high in Case 2, and below average in Case 3.

The lack of correlation between the degree of hydrocephalus and the intelligence was noticeable in Case 2.

As Jansen stressed, there are degrees of achondroplasia and slight examples of the disease may not be recognized. It is conceivable that the chondrodystrophic disturbance may at times be almost confined to the base of the skull. Hydrocephalus may then develop and the essential fact of achondroplasia may pass undetected.

Summary

The neurological complications which arose in three achondroplasic adults are described. Internal hydrocephalus was present in each case and probably resulted from the characteristic shortness of the base of the skull. There was no evidence of platybasia nor of any deformity of the foramen magnum.

Two of the patients developed severe cauda equina compression as a result of multiple massive lumbar intervertebral disc protrusions. At operation it was also observed that the spinal canal was generally narrowed. The achondroplasic father of one of these cases had also developed paraplegia requiring laminectomy. The third patient had a spastic tetraplegia with subluxation of the second cervical vertebra upon the third. There was spinal manometric block when the neck was slightly flexed or extended. No bony exostosis could be demonstrated in the upper cervical level.

In Cases 1 and 2 the bodies of the cervical vertebrae were tall. In Case 3 all the vertebral bodies were shallow and broad. In all three cases the posterior surfaces of the bodies of the lumbar vertebrae were concave and the intervertebral disc spaces were unusually wide. Similar radiological signs were observed in varying degree in five out of six other achondroplasic adults.

None of the three patients with spinal complications had ever been engaged in tumbling or acrobatics.

I am grateful to Professor Lambert Rogers, Mr. Charles Langmaid, and Dr. Geoffrey H. T. Lloyd for their observations and assistance, and to Dr. Arthur M. Jones for the radiographs. Clinical photographs were taken by Mr. H. A. Griffiths, of the Department of Clinical Photography, Llandough Hospital.

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J. Neurol. Neurosurg. Psychiat., 1952, **15**, 253.

A SEX-LINKED VARIANT OF GARGOYLISM

BY

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In the condition first described by Hunter (1916–17) and named " gargoylism " by Ellis, Sheldon, and Capon (1936), opacity of the corneae is, with mental deficiency, osteo-chondro-dystrophic changes, and hepatosplenomegaly, cited as a cardinal sign. That it is not an invariable component of the syndrome has since become evident from the reports of De Lange and Woltring (1936–37), Ross, Hawke, and Brown (1941), De Lange, Gerlings, de Kleyn, and Lettinga (1943–44), Lurie and Levy (1944), Debré, Marie, and Thieffry (1946), and Njå (1945–46) who describe cases typical in other respects but lacking corneal opacity.

The genetic aspect of gargoylism was investigated by Halperin and Curtis (1942). In reaching the conclusion that the condition is " probably due to the action of a single recessive autosomal gene " these authors, however, make no distinction between cases with corneal opacities and those without. The first indication that there may be a specific variant of gargoylism characterized by clear corneae and a sex-linked heredity appears in a paper by Wolff (1942). Writing from the otological standpoint, she gives the pedigree of a family in America which included four cases, apparently sex-linked, in three generations. One of these is described in some detail, but although lenticular anomalies were noted in the right eye and an abnormality in the disc in the left, the corneae are not specifically mentioned. It is not unreasonable, therefore, to suppose that they were clear.

In 1945–46 Njå described five cases, again without corneal opacities, in a Norwegian family. The pedigree was so strongly suggestive of a sex-linked heredity that this author concluded that it was justifiable to set up a special type, " a sex-linked recessive form where the corneae are clear ". This paper describes a group of cases in a family in England which would appear to support this view.

Clinical Report

The pedigree of our English family is shown in Fig. 1. Nothing is known of the original parents (i, 1 and 2) except that the father was a seafaring man who died " of a stricture " at 47 years and the mother " one of 13 who were all well ". There is no evidence of consanguinity.

As in all previously described gargoyles with clear corneae the sex of our cases was male. No female members of the family exhibited the trait.

The significance of the symbols in Fig. 1 shown in solid black is as follows: Four (ii, 1 and 7; iii, 13; and iv, 6) are undoubted cases of the condition under discussion. One (iv, 13) is almost certainly another case though confirmation is lacking. In the remaining case (iii, 7) there can be no more than a suspicion. Little is known of this child and nothing is known of the siblings (iii, 6) or of the descendants, if any, as the family had lost touch with the mother many years ago. The suspicion arises, as the child is said to have been " not quite right ", and it is said that his death at 7 years was a " happy release ".

The first five of these cases will be described in degrees of detail which vary according to the information available.

Case 1.—This boy (Fig. 1, iv, 6) was known to one of us (C.G.M.) when 7½ years old and had been under his care from age 11½ until death at 13 years and 7 months.

The earlier history was that the mother's pregnancy and labour were normal and the child's birth weight 3·85 kg. Mental deficiency was not suspected until towards the end of the first year. The child was unable to sit up at 12 months but walked at 18 months and talked a little at 2 years. Dentition had started at 8 months. At 2 years and 9 months the head circumference was 55 cm., the vertex was high, and the hair coarse. Teeth were rather widely spaced. The back was flat and rigid in appearance when the patient walked. The costal margin was flared and there was a Harrison's sulcus. The liver was palpable but the spleen was not. In the lateral view a radiograph of the spine showed some vertebrae to be ovoid and some to have the hooked processes characteristic of gargoylism.

At 7½ years the child weighed 19·73 kg. and his height was 99 cm. There was no true kyphosis but he walked stiffly, flexed a little at the hips. The head was large

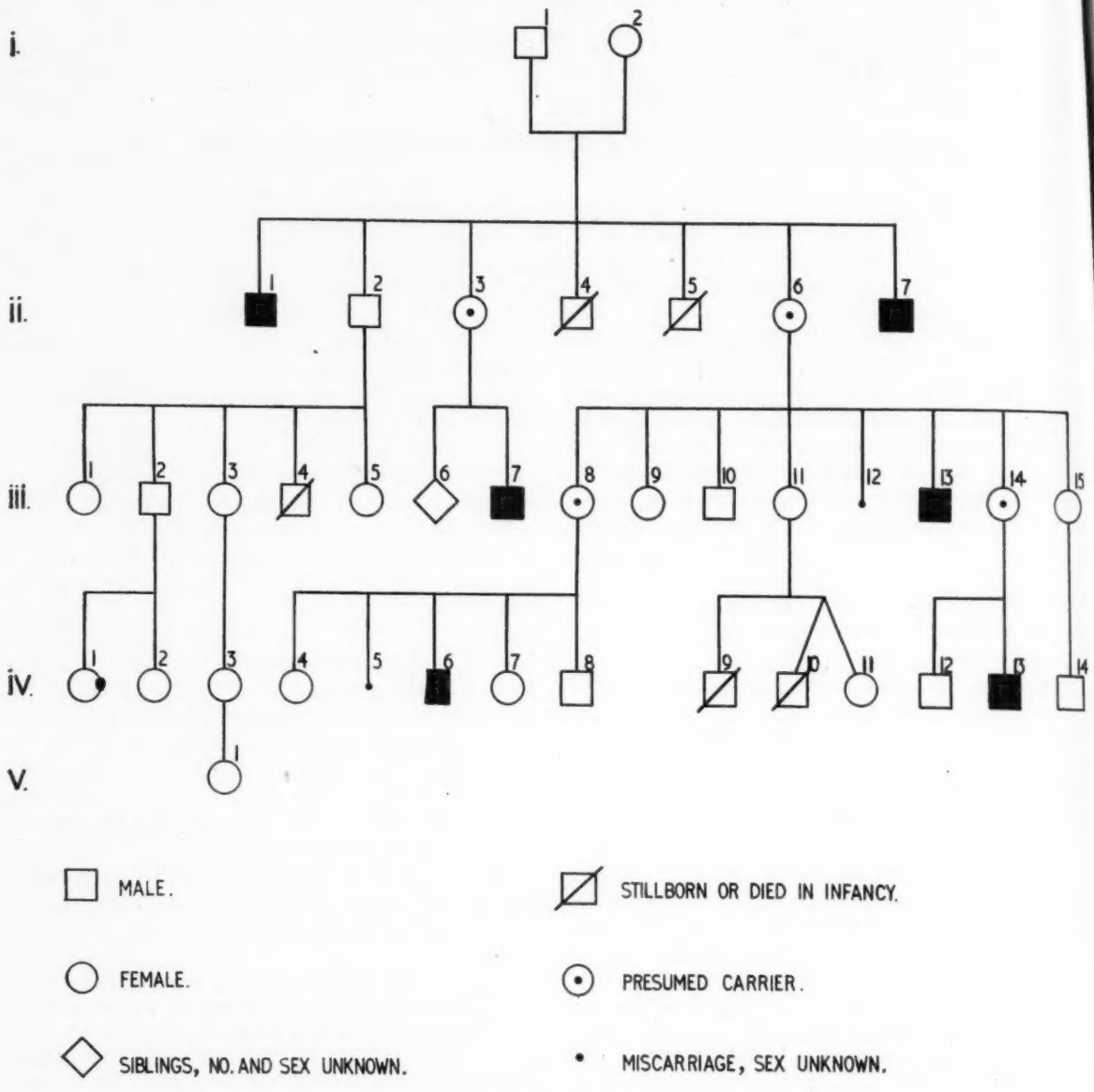

FIG. 1.—Pedigree of affected family.

with frontal bossing; there was a narrow, arched palate and a broad nose with slight depression of the bridge. The ears were placed low. Teeth were healthy but $\frac{6|6}{6|6}$ had not yet erupted. The features were heavy and respiration was snuffly. The hair was coarse, the eyebrows bushy, and there was a mild generalized ichthyosis. The liver was enlarged and the abdomen protuberant. There were umbilical and right inguinal herniae and a left hydrocele. There was a loud systolic murmur at the apex of the heart. No corneal opacity was present.

Mentally, at 7½ years the child remained at a stage of development barely equivalent to the 2-year level; he could say a few words, feed himself, and was clean in habits but he played like a baby.

At 12½ years deafness was noted but the patient resisted all attempts at otological examination. At this

time his respiration was noisy and he suffered from a chronic catarrh of the upper respiratory tract. By 13 years a long-standing increase of intracranial pressure had led to some swelling of the optic discs and blurring of their edges. The child's appearance at this age is shown in Fig. 2. By this time he was in a play group at the Cell Barnes Colony school where his histrionic abilities earned him the nickname "Inspector". He was unable to do anything constructive but was fond of giving his imitation of a person writing fluently.

At 13 years and 7 months the patient died suddenly from a combination of causes to be described below.

Case 2.—His disease (Fig. 1, iii, 13) was not diagnosed during life but has been adequately described by his mother and by his sister (the mother of Case 1). Medical notes and a photograph are also available.

That the mental level of this case was rather higher than that of Case 1 is evident from the history. Backwardness was not suspected at such an early age, and at 11 years the child played in a fashion with other children, knew two or three letters, could write a little and be occupied constructively. After leaving a special school at 16 he could help a little in the home and run errands.

Records provide the following information on the physical aspect of the case: At 11 years his height was 120 cm. and the head circumference 54·5 cm. He held his neck very stiffly and was very deaf. By 16 years the child was said to be suffering from heart disease and from "a crippling defect of the fingers". Eighteen months later this was diagnosed as "multiple arthritis of the small joints" and the hips were also becoming involved. Presumably the "arthritis" was the characteristic osseous dystrophy.

An ophthalmologist's report at 18½ years gives the retinoscopic findings as +9·5 D in each meridian in both eyes and, as no mention is made of the corneae, these are presumed to have been clear.

This patient died suddenly at 21 years of heart trouble. A necropsy is said to have been performed but no records can be traced.

Case 3.—From his sister's description and from the family album we learn that this boy (Fig. 1, ii, 1) was a little taller than Case 1 and had no kyphosis. He had a very large head, "black hair like wire", bushy eyebrows, thick lips, a prominent abdomen, and "no neck; his head rested on his shoulders". On enquiring as to corneal opacities we are told that he had "beautiful eyes". His mental level was such that he "never went out of doors, used to run about with the broom, was sometimes bad-tempered and would throw things". It is said that he enjoyed looking at newspapers but was almost certainly unable to read. He died at 25 years of dropsy after a month's illness.

Case 4.—From the same sources as in the previous case we learn that this child (Fig. 1, ii, 7) was very small and, at the time of his death from bronchopneumonia at 16 years, was about 91 cm. in height and had a head which, though large, was not as big as his brother's (Case 3), and a "big stomach". He "always seemed to have a cold". He had bushy eyebrows but fair hair of normal texture. Enquiry as to his eyes elicited the information that they were "sparkling". He went to school until he was 14 but never learned to read or write. He was very simple and of a "sweet disposition".

Case 5.—At present there can be no more than a suspicion regarding this case (Fig. 1, iv, 13) as permission to examine the child has been refused. However, a hospital report at 1 year stated "... keeps his mouth open a lot and sleeps with an open mouth. Tonsils ++. Appearance rather suggests a mongol but there is no supporting evidence for it other than facial expression ... Harrison's sulcus marked. Sternum indrawn. Frontal bossing. Bridge of nose satisfactory. *X*-ray skull n.a.d." At 14 months a note stated "does not walk yet but crawls and would not stand alone". Suspicion in this case rests on the evidence of osseous abnormalities, unusual appearance, retarded development and, above all, on the opinion of the mother of Case 1 who states that, at 4 years, there is a strong resemblance to her own affected child at the same age.

Necropsy Findings in Case 1

Necropsy was performed 42 hours after death. The body was that of a stunted boy (height 107·5 cm.: weight 25·5 kg.) of fair muscular development and normal proportions except for a large head and short neck. The bridge of the nose was sunken but there was no prominence of the supra-orbital ridges nor of the mandible. The corneae showed no opacity. There was no kyphosis and no deformity of the hands or feet.

In addition to a protuberant abdomen there was a small empty umbilical hernia (length of sac 1·5 cm.; orifice 0·9 cm. diam.). The external genitalia were normally developed.

FIG. 2.—Case 1 at 13 years.

Skeleton.—The skull was considerably enlarged (circumference 57·0 cm.; diameters, fronto-occipital 19·0 cm., bi-parietal 14·0, occipito-mental 23·0 cm.), and moderately thickened and there was considerable bulging of the orbital roofs into the cranial cavity. No other bony projections into the cranial cavity from the skull base were present. The pituitary fossa was greatly enlarged (1·9 cm. antero-posteriorly, 1·7 cm. from

side-to-side, 1·4 cm. deep). Both middle ears contained a small amount of greyish, viscid liquid.

Bilateral grooving on the lower thoracic wall was due to indentation at the costochondral joints of ribs 7 to 9. There was no thickening of the costochondral junctions.

Some reduction of the normal curvatures of the spine in both thoracic and lumbar regions was evident.

Nervous System.—No abnormality of the dura, which was tense, was found but there was white opacity and prominent thickening of the leptomeninges over the superolateral surfaces of the cerebrum, especially in the neighbourhood of the superior longitudinal venous sinus. The subarachnoid space was distended by clear, pale yellowish liquid. Internal hydrocephalus was pronounced. An enlarged, non-cystic hypophysis filled the pituitary fossa.

Cardiovascular System.—The heart weighed 160·0 g. Its chambers were of normal proportions and the myocardium, which was dark red, had the following maximum thicknesses in the different chambers: left ventricle 1·1 cm., right ventricle 0·4 cm., left atrium 0·3 cm., and right atrium 0·2 cm. All cusps of the aortic and mitral valves were uniformly thickened (0·2 cm.), rigid, and opaque white. Their surfaces were smooth. The coronary arteries were fully patent and free of atheroma. The foramen ovale was closed. There were a few small opaque yellow flecks of atheroma in the abdominal aorta. Medial to the junction of the left internal jugular and subclavian veins was a multilocular cystic mass (3·3 × 1·8 × 1·2 cm.) containing fresh and partly clotted blood. It was embedded in fat, through which ran small veins communicating with the internal jugular vein.

Respiratory System.—In the naso-pharynx congested polypoid mucosal projections greatly reduced the lumen, and lymphoid tissue in the pharynx was prominent. The palatine tonsils were greatly enlarged and there was considerable narrowing of the glottis caused by oedema of the ary-epiglottic folds.

The upper lobes of the lungs were bound to the thoracic walls by fibrous adhesions. There was uniform congestion of both lungs with thick, greyish mucopus in the bronchi of all lobes.

Lymph Nodes.—One upper deep cervical lymph node on each side was enlarged (larger 3·3 × 1·8 × 1·2 cm.) and had soft, greyish-white cut surfaces. All other lymph nodes appeared normal.

Alimentary System.—The stomach, grossly distended by greyish liquid, occupied about one-third of the available space in the abdominal cavity. The mucosal surface of its thin wall was closely studded with slightly projecting, miliary and sub-miliary, pale brownish nodules.

There was considerable enlargement of the liver 1,270·0 g.) which in the mid-line extended 11·6 cm. beyond the tip of the xiphoid process, and on the right side 8·0 cm. beyond the costal margin. It was paler than normal, the capsule was tense, and cut surfaces showed very numerous ill-defined, pinkish-grey and opaque white foci of up to 0·3 cm. diameter.

Spleen.—Though enlarged (13·0 × 8·4 × 3·5 cm.; 197·0 g.), the spleen was not as large as its position, lower margin 6·0 cm. beyond the costal margin, suggested. This was due partly to displacement by the greatly distended stomach. It was firm and had a thin, smooth-surfaced capsule. On cut surfaces fine trabeculae were distinct but Malpighian bodies were not recognizable, being lost in the pale greyish-brown pulp.

The thymus was enlarged (10·0 × 6·5 × up to 1·9 cm. thick; 33·0 g.). No abnormality was found in the adrenals (together 8·7 g.), thyroid (20·0 g.), pancreas, kidneys (left 79·0, right 67·0 g.), ureters, bladder, prostate, and testes (each 3·0 g.).

The sudden death was thought to be due to a combination of cardiac and respiratory failure.

Histological Findings in Case 1

Spleen.—There was an appreciable amount of diffuse cellular fibrosis: otherwise no special features. No sudanophil material was stainable, no doubly refractile lipoid was present, and no vacuolated cells could be found.

Liver.—Complete derangement of lobular pattern was due to swelling of the hepatic cells, which greatly compressed the sinusoids, and were thrown out of their normal arrangement in columns. The liver cells had thick cytoplasmic walls enclosing pale, sparsely granular cytoplasm in which only small numbers of fine vacuoles were recognizable (Fig. 3). These cells, with Best's carmine stain, gave only a faint positive reaction, indicating that little glycogen was present: it was the granular material which stained positively. Sudan stains for neutral fat showed only small widely scattered groups of small globules. Examined under polarized light, no doubly refractile lipoid could be found. Portal tracts contained small foci of lymphocytic and neutrophilic polymorph infiltration and occasional small groups of multinucleate giant cells.

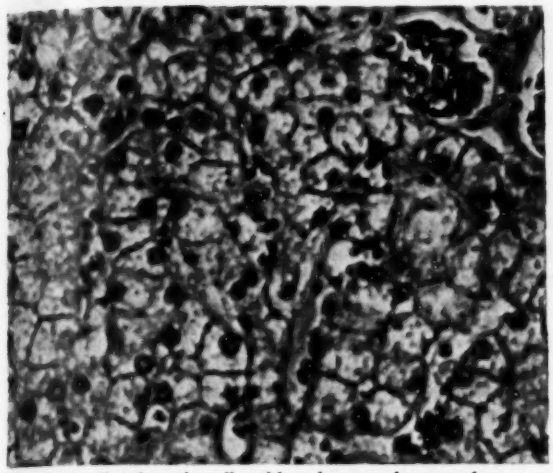

FIG. 3.—Swollen hepatic cells with pale, granular cytoplasm compressing and obliterating sinusoids. × 320.

Costochondral Junction.—There was an almost completely static straight line with practically no proliferation of cartilage, a very narrow zone of provisional calcification of cartilage, and little osteoblastic or osteoclastic activity (Fig. 4). A striking change was the presence of a thick layer of pale-staining, fibrous tissue between the thick periosteum and the irregular bony corticalis of the

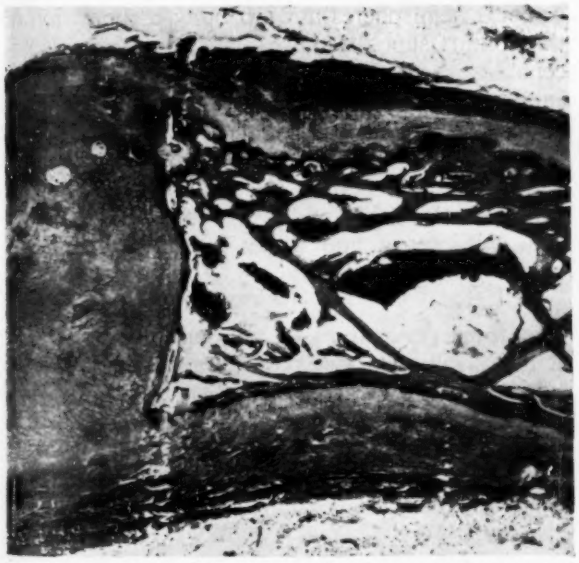

FIG. 4.—Costochondral junction showing almost complete arrest of endochondral ossification and thickened periosteum. × 7.

rib. The collagenous fibrillae in this layer ran obliquely between the corticalis and the periosteum and were separated by fairly numerous, irregularly scattered clear cells, in the cytoplasm of some of which was a small amount of pale-staining granular material (Fig. 5). Clear cells of the same type, though less numerous, were present throughout the periosteum. In sections treated with Sudan III, Sudan black B, and by the periodic-acid-Schiff method these pale cells failed to stain.

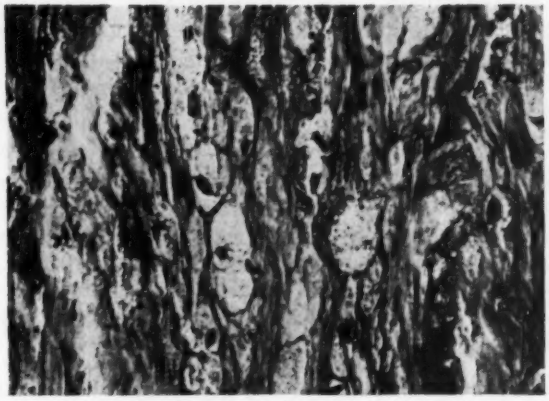

FIG. 5.—"Clear" cells scattered through the thickened periosteum of a rib. × 320.

Pituitary.—Enlargement was due to an increase in size of the anterior lobe where there was a paucity of eosinophilic cells and a relative increase in the number of cells with basophilic granules. The commonest cell was a degenerate one with finely vacuolated pale cytoplasm which appeared to be derived from the basophilic cells.

Heart.—Thickening of the mitral cusps was due to dense, poorly cellular fibrous tissue. Much of the collagen stained poorly by van Gieson's method. Between the collagen fibres there were fairly numerous, pale, ovoid cells with small, usually pyknotic nuclei and finely vacuolated or granular cytoplasm (Fig. 6). They were identical in appearance with the clear cells in and below the periosteum of the ribs.

There was no abnormality in the myocardium.

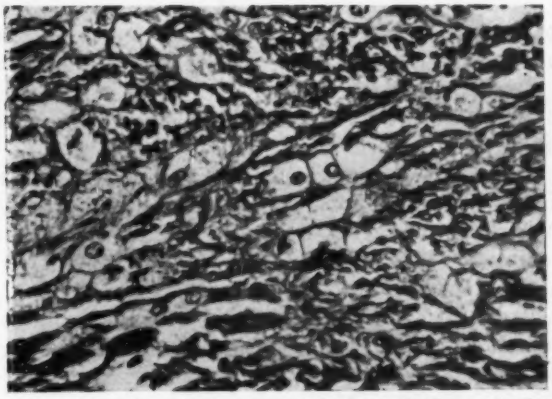

FIG. 6.—Separation of collagenous tissue in a mitral cusp by large "clear" cells. × 320.

Lymphoid Tissue.—The polypoid mucosal thickenings of the naso-pharynx, the palatine tonsils, and the upper deep cervical lymph nodes all showed great lymphoid hyperplasia and subacute inflammatory changes.

The fine nodularity in the gastric mucosa was due to the presence of lymphoid follicles.

Eye.—No abnormality was present.

Left Supraclavicular Tumour.—This consisted of a *congeries* of large vascular spaces containing blood and embedded in fibrous and adipose tissue in which there were small lymph nodes. Acute inflammatory changes were superimposed.

Lung.—Long-standing chronic bronchitis and acute congestion were present.

Thymus.—Glandular thymus was of normal structure except for collections of neutrophilic polymorphs in and about Hassall's corpuscles.

No significant abnormality was present in sections of the kidneys, prostate, testes, thyroid, parathyroids (two examined), and adrenals.

Unfortunately mishap overtook the stored brain, vertebrae, and long bones, so that these were not available for further examination

Discussion

Published accounts of necropsy findings in cases of gargoylism are few. Strauss (1948), in a complete review of the literature, refers to descriptions of the pathological changes in 15 cases but notes that several are incomplete and some are doubtful instances of gargoylism. She adds a new case, and Russell (1949) in her monograph on hydrocephalus mentions another. Of these 17 gargoyles only two were of the type with clear corneae (De Lange and others, 1943–44 ; Njå, 1945–46).

While we are unable to give a complete description of the pathological changes in Case 1, our findings allow confirmation of the clinical diagnosis of gargoylism and raise points of interest. Because of Strauss's recent review, only a few of these will be referred to briefly.

With the material available, it was not possible to demonstrate the presence of lipoid, though large, pale cells with vacuolated finely granular cytoplasm, resembling those illustrated by Strauss, were present in the thickened cusps of the mitral valve and in the widened periosteum of a rib. Cytoplasmic changes of similar appearance were present in the distended hepatic cells but in none of the other organs examined microscopically. The cornea and other parts of the eye were normal.

The skeletal abnormalities in our case resemble those described in other gargoyles, and the histological finding of arrest of ossification at the costochondral junctions with periosteal thickening and infiltration by vacuolated cells is very similar to that of Strauss. Though no differential count of the cells of the anterior pituitary was made, the impression was gained from examination of sections that there was a reduction in the number of cells with eosinophil granules. Till this is confirmed by other necropsy studies its possible relationship to the characteristic dwarfism of gargoyles can be only tentatively suggested.

Thickening and rigidity of the cusps of the mitral and aortic valves have been described previously, and our findings and views are very similar to those of Strauss who regards the change as due not to endocarditis but to the accumulation of vacuolated cells and fibrosis. The common occurrence of sudden death due to heart failure is probably related to these valvular lesions.

There is in our case no support for Strauss's opinion that when hydrocephalus occurs in gargoyles it is due to compression of the brain stem by bony elevations at the base of the skull. As in Russell's case, ours showed thickening of the leptomeninges over the superior surfaces of the cerebral hemispheres and no bony compression of the brain stem. Her explanation that hydrocephalus results from this leptomeningeal fibrosis is supported by our findings but we were unfortunately unable to examine the meninges histologically for the lipoidal material which she describes within the arachnoid cells. Njå's Case 2, which had clear corneae, also showed hydrocephalus associated with " thickened and milky " leptomeninges.

Summary

In three generations of an English family, four and possibly five cases of gargoylism, in which corneal opacity was lacking, are described. All five subjects were males and they appeared to inherit the condition as a sex-linked recessive character.

The necropsy findings in one of these cases are described.

We wish to express our gratitude to Dr. Noel H. M. Burke, Medical Superintendent, Cell Barnes Colony, for permission to publish the details of Case 1, and for his encouragement and assistance. We are indebted to Dr. A. Shapiro for calling our attention to the families described by Wolff and by Njå, whose reports we had not yet seen when our own investigations were finished. To Professor L. S. Penrose we extend our thanks for his encouragement and advice on the genetic aspect of the

work. We are indebted to Dr. W. G. Wyllie for notes on the earlier history of Case 1 and to Dr. W. Templeton for giving access to the notes on Case 2. We also wish to thank Mr. A. E. Reynolds for the photograph and Mr. L. A. Cowles for the photomicrographs. Finally we would place on record, with gratitude, the help given by many members of the family we have described.

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Addendum

Since dispatching our paper we have received from Dr. Ivor Balfour the following further details of Case 5 which he examined at 5 years and considered to be one of gargoylism :

Height 99 cm.; weight 16·4 kg.; head circumference 52 cm. Marked supra-orbital ridges. Broad alae nasi and nasal discharge.

Ears low on head. Thick neck. ? early kyphosis. Genu valgum. Broad hands ; terminal phalanges cannot be extended, i.e. claw fingers. No corneal opacities. Apparently some mental retardation.

A radiograph showed considerable enlargement of the sella turcica, especially the antero-posterior measurement; some thickening of bones on either side of the coronal sutures, but no evidence of the other characteristic bony changes reported in the literature.

Abdomen was distended ; umbilical hernia. Liver and spleen enlarged and easily palpable.

Blood cholesterol 140 mg.%. W.R. negative. White blood count 4,400, and 3,300 on the two occasions tested. Giemsa-Wright stain (Reilly method) showed no pink granules.

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ELECTROENCEPHALOGRAPHIC STUDIES IN CEREBRAL ANGIOMA

BY

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An enquiry into the value of electroencephalography (E.E.G.) in cases of cerebral angioma was prompted by the need for accurate information with which to aid E.E.G. reporting. There are few references in the literature to which one can turn, and these are mainly isolated studies of cases of the Sturge-Weber syndrome.

In the present investigation a brief description of the clinical material used is followed by an analysis of the E.E.G. results. All figures given in percentages or years are changed to the nearest whole numbers for simplification.

Technique

Six-channel machines with amplifier performance complying with the E.E.G. Society recommendations (one Ediswan Mark 1, and two machines built in the National Hospital) were used in all cases. The scalp electrodes were of the pad variety, i.e. 1 cm. diameter pads dipped in 5% saline and attached with silver screws to moulded perspex bases, as described by Grey Walter (1950). Nineteen electrodes were used in standard positions in antero-posterior or transverse great circle lines at a distance of 4 to 7 cm. from one another, and were held attached to the scalp by a rubber harness fastened under the chin. Seven or eight standard positions were adopted in all cases and the recordings were bipolar without exception. Provocative techniques were used where indicated and overbreathing was done as a routine except where the condition of the patient rendered it inadvisable.

The routine time constant was 0·3 seconds and the high frequency response was 15% down at 75 cycles per second (c/s).

Clinical Material

The material comprised 55 consecutive cases of cerebral angioma proved by arteriography which were admitted to the National Hospital during the five and a half years from late 1946 to early 1952. Cases suspected but not proved, and cases verified but not investigated by the E.E.G., were excluded from the series. Of the 55 cases, the pre-operative record of one was unsatisfactory while in two cases only the post-operative records were available. These three records were excluded from the analysis of results before operation. A total of 94 records was made.

Clinical Analysis

Of the 55 patients, 27 were male and 28 female. Their age composition is shown in Table I. The duration of symptoms before E.E.G. examination was on the average seven years. Brain (1951) states that a long history is common, and our figure shows an interesting comparison with that obtained in this laboratory for the duration of symptoms in cases of intracranial aneurysms which was approximately three years, and that for spontaneous subarachnoid haemorrhage which was roughly eight months before E.E.G. examination was carried out.

The angioma was right-sided in 25 cases, and in 30 was left-sided. Twenty-three were parietal, seven temporal, six occipital, seven frontal, five parieto-frontal, five parieto-temporal, one parieto-occipital, and one occipito-temporal. Over half were supplied by the middle cerebral artery. The remainder were supplied, in order of frequency, by the anterior cerebral, posterior cerebral, and the vertebral arteries. Four involved the carotid syphon.

Thirty-six patients presented with epilepsy or developed fits during the course of the illness, a complication which is common, as Brain (1951) points out, while 17 had an associated mono- or hemi-paresis. Subarachnoid haemorrhage had occurred in 18 cases (with a further two doubtful ones). Cushing and Bailey (1928) and others do not mention what percentage of angiomata is complicated by haemorrhage but merely state that when subarachnoid bleeding supervenes death commonly results. The average age of patients with subarachnoid haemorrhage before examination was 34 years with a maximum incidence in the third and fourth decades. This age grouping is very similar to that of the whole series (Table I). In addition, it may be mentioned that only 13 patients complained of severe headache when seen. Twenty-one had an audible intracranial bruit. In two patients the angiomata bled into the brain, and one of these died (both grade 3, see later).

TABLE I

THE AGES OF THE 55 PATIENTS

Age (in decades)	Male	Female	Total (55)
Second ..	4	5	9
Third ..	8	6	14
Fourth ..	5	7	12
Fifth	4	9	13
Sixth	6	1	7
Average age	31 years	34 years	32·5 years

Craniotomy was performed in 28 cases (51%) where the angioma was superficial or partly so. In two of these the operation was terminated without any surgical interference with the angioma being attempted. The angioma was removed in 19 cases and its vessels ligated within the skull in seven. Four patients had the carotid artery on one or other side compressed under E.E.G. control before operation, three of these being included in the total of five patients whose carotid artery was later ligated. No E.E.G. changes were recorded in these cases during carotid compression for 20 minutes. The remaining 22 patients were considered to be unsuitable for operation. Two patients died after operation.

E.E.G. Analysis

The cases are divided into three groups according to the E.E.G. analysis (Table II). Group 1 includes normal or doubtfully abnormal records. Group 2 comprises those records showing mild unilateral abnormalities such as minor aysmmetries of amplitude or 4–7 c/s activity. Group 3 consists of records showing severe unilateral abnormalities, e.g. marked asymmetry between the two hemispheres, high voltage slow wave foci, or gross disorganization of the normal rhythms. These divisions are naturally somewhat arbitrary.

In Group 3 one case showed lateralizing, and one case localizing, features on the side of the brain opposite to the angioma. A third, showing a high voltage slow wave focus, had an intracerebral haemorrhage in addition to the angioma. Two patients had an aneurysm as well as an angioma, the one having the record which was unsatisfactory, the other showing an E.E.G. grading (Table II) before operation of 2 (focal), and 13 months after operation of 2 (local). The E.E.G.s of another two patients when first recorded were normal, and during serial readings progressed to group 3, one focal and one local. It may be of interest to add that one of the two patients last mentioned was examined five times before operation over the course of a year, the last four records showing a persistent slow wave focus in the left occipito-temporal area. As he suffered only from epilepsy and showed no physical signs, he was not admitted to hospital till he had been under observation for a year when an angioma was found in the position located by the E.E.G. Two other patients showed localized 8 c/s activity distinct from the normal alpha rhythm over the site of the lesion, both of which were in the left posterior parietal position.

TABLE II

CLASSIFICATION OF CASES ACCORDING TO DEGREE OF E.E.G. ABNORMALITY IN 52 CASES OF ANGIOMA

Group	1	2	3	Total
Number of cases	17	18	17	52
No lateralizing features	17	—	—	17
Lateralizing features only	—	7	4	11
Lateralizing and localizing features ..	—	5	4	9
Focal features	—	6	9	15

Table II shows that out of 52 patients, 17 (33%), i.e. Group 1, give normal or doubtfully abnormal records, while the E.E.G.s of the other 35 cases (67%) show lateralizing or localizing features. This last figure may be compared to an approximate one of 37% for intracranial aneurysms found in this laboratory when similar criteria are used.

A more detailed analysis of the E.E.G. findings before operation shows that alpha (8–13 c/s) activity is completely absent in three records, is present on one side only in four, and is asymmetrical in 17. The voltage is lower on the same side as the lesion in 13, and is greater on the same side in four, there being no apparent reason for this difference. Theta (4–7 c/s) activity is present in 34 records, mostly non-focal, usually irregular and confined to the side of the lesion, and predominantly at 6–7 c/s. The voltage is mostly low. Delta ($\frac{1}{2}$–$3\frac{1}{2}$ c/s) activity is present in 23 records, mostly focal and of moderate to high voltage, and usually at 2–3 c/s. Beta (14–30 c/s) activity is seen in the records of 13 patients, mostly small in amount and frontal and bilateral, sometimes spread over the whole cortex, and unilateral in one case over the site of the lesion. Eleven of these patients suffered from epilepsy, and the remaining two were not receiving medication.

A composite picture might therefore show a low to moderate voltage alpha rhythm, possibly asymmetrical, irregular 6–7 c/s activity on the side of the lesion, fairly localized, with perhaps a high voltage delta focus on the same side as well.

Post-operative records of eight patients were available, of whom six were examined before operation. The angioma was removed in five cases and its vessels ligated in one. Two patients had the internal or common carotid artery on one side ligated. Two patients within the six weeks after operation showed a more abnormal E.E.G. than before. All records up to seven years after operation were still abnormal, the E.E.G. gradings being almost unchanged as compared with those taken before operation, both for craniotomy and carotid ligation, with the possible exception that the abnormal slow activity was less strictly localized. On the other hand, no definite correlation between the clinical state and the E.E.G. state was found, as six of the eight patients showed clinical improvement, a finding not supported by the E.E.G.

When the E.E.G. abnormality is correlated with age and the duration of symptoms before E.E.G. examination it is seen that with decreasing age and duration of symptoms the E.E.G. is progressively more abnormal (Table III). This general trend is seen in other disorders. Fits have little bearing on these E.E.G. changes, as of the 34 patients with records taken before operation and giving a history of epilepsy, only four showed definite epileptic activity (spikes) on their records, with a further six showing doubtful dysrrhythmic activity. These last 10 patients are distributed fairly evenly among the three E.E.G. groups. (As nearly all the 34 patients are on anticonvulsant therapy, fast activity is not considered here as an epileptic manifestation.)

TABLE III

CORRELATION OF E.E.G. ABNORMALITY WITH AGE AND DURATION OF SYMPTOMS AT TIME OF E.E.G. EXAMINATION

E.E.G. grading (see Table II)	1	2	3
Number of cases	17	18	17
Average age (years)	37	30	30
Duration of symptoms (years)	11	6	4

Subarachnoid haemorrhage too occurs almost equally among the three E.E.G. groups (five, eight, and five cases respectively), but it is of interest to find that when 18 cases of angioma with subarachnoid haemorrhage, taken from this series, are compared with the series as a whole, the percentage of E.E.G. lateralizing abnormality rises slightly from 67 for the whole to 72 for the selected group.

An equivalent group of 18 cases of subarachnoid haemorrhage derived from confirmed cerebral aneurysm gives lateralizing E.E.G. abnormalities in 50% of the cases. This suggests that a case of subarachnoid haemorrhage showing E.E.G. lateralizing abnormality is more likely to be due to an underlying angioma than to an aneurysm, though this obviously does not apply to the individual case.

It is known (e.g., Roseman, Bloor, and Schmidt, 1951) that the sooner after a subarachnoid haemorrhage the E.E.G. is taken, the greater is the likelihood of its being abnormal. Conversely, should the patient live and not have an intracerebral haematoma, the E.E.G. improves with time.

Discussion

Broager and Hertz (1949), using the bipolar method on one patient, found a left-sided parasagittal premotor focus with constant phase reversal before operation, and diffuse slow activity one week after operation. Raskin (1949) described slowing of 7–8 c/s activity in all leads in another. Radermecker (1951) reviewed nine patients with facial naevi, six of whom had calcified cerebral lesions. The E.E.G. findings in this group did not show any common pattern. Other cases are described by Cornil, Paillas, and Gastaut (1949), by Monnier and Mutrux (1949), and by Craig (1949). Larmande (1948) quotes Green's description of an infant with epilepsy who showed high voltage $2\frac{1}{2}$–6 c/s activity with an area of non-activity over the region of the angioma. Gastaut and Bonnal (1952) describe a case of angioma in the right temporal lobe, confirmed by arteriography, with focal $2\frac{1}{2}$ c/s waves over the site of the lesion.

The relation of the E.E.G. abnormality to the time factor in cases of subarachnoid haemorrhage, irrespective of the underlying pathology, is stressed by Greenstein and Strauss (1946) and by Roseman, Bloor, and Schmidt (1951). Bailey (1948) points out that though epilepsy is commonly associated with angioma it is in no way specific, and in this series E.E.G. confirmation of epilepsy was obtained in only a small proportion of those patients giving a history of it. The association of epilepsy with fast activity on the records in this series cannot be assessed accurately as most of the patients suffering from epilepsy are taking barbiturates.

Two patients show progressively deteriorating records with advancing clinical changes. Another two show more severe E.E.G. abnormalities within six weeks of operation than before. This is in agreement with other forms of pathology. But no correlation is found between the post-operative clinical and post-operative E.E.G. states. It is not known what significance to attach to the two records where localized 8 c/s activity is found over the site of the lesion.

In the few cases where carotid compression on the appropriate side was done under E.E.G. control before operation no electrical changes were recorded. The same negative result was obtained in the majority of cases of a much larger group of intracranial aneurysms tested in this laboratory in the same way.

The left ventricle of the heart of one patient in the series was found to be enlarged, though no cardiac disease or hypertension could be demonstrated. This raised the possibility that the angioma was causing an abnormal circulatory state sufficient to produce the cardiac enlargement. From an analysis of samples of blood taken from the patient's heart and internal jugular vein it was estimated that the cardiac output was increased by 40% (7·2 litres per minute) above the normal for the patient (5·1 litres per minute), and that the circulation through the brain was approximately doubled, giving an arteriovenous shunt of the order of one litre per minute. Gibbs, Maxwell, and Gibbs (1947) state that the average resting blood flow in the brains of seven patients (not suffering from angioma) is 617 ml. per minute.

To sum up, no specific E.E.G. disturbance diagnostic of angioma could be seen in the 94 records of the 55 patients. In general, an angioma gives a similar E.E.G. picture to that of cerebral tumour which characteristically shows localized high voltage irregular delta waves. However, the impression is that with angioma raised intracranial pressure and the spread of oedema play on the whole a lesser part than in cases of tumour, giving less generalized delta activity that will obscure a true focus, and allowing a clearer localization of the intermediate slow activity. The same distinction serves to differentiate angioma from cerebral abscess, where in addition, the slow delta waves at ½–2 c/s are usual. But as Cobb (1950) points out, the determination of pathology by the E.E.G. is at the most, uncertain.

Summary

The electroencephalograms of 55 cases of proved cerebral angioma have been analysed.

The E.E.G. gives a lateralizing or localizing indication in 67% of the cases, the usual findings being localized 6–7 c/s activity and/or high voltage 2–3 c/s waves showing phase reversal over the site of the lesion. These changes, with minor variations, are usually seen with other forms of cerebral tumour.

The value of electroencephalography as a diagnostic aid in angioma is discussed.

I am indebted to Dr. W. A. Cobb for his advice and criticism in the preparation of this paper, and to the physicians and surgeons of the National Hospital for access to their case records.

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DISORDERS OF PERCEPTION AND PERFORMANCE IN A CASE OF RIGHT-SIDED CEREBRAL THROMBOSIS

BY

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The disorders of perception and performance which accompany cerebral injury have a particular fascination in neurology, in that they cannot always be explained solely in terms of the sensory loss or motor deficit observed.

It has been known for some time that in right-handed persons lesions of the right cerebral hemisphere involving the fronto-parietal region may produce, in addition to left hemiplegia and left hemianaesthesia, an agnosia, a failure to recognize the left side, or an anosognosia, a failure to recognize disease of the paralysed side. Typical cases have recently been reported by Sandifer (1946) and by Roth (1949). It has also been shown that right parietal lesions may result in a corresponding neglect of external space on the hemiplegic side (Brain, 1941). There is some doubt as to whether this is a defect characteristic of cerebral lesions on the right side only, or whether it is present but masked by grosser disabilities after left-sided parietal lesions (Brain, 1945). There may also be a failure to solve visual problems and to perform constructional tasks involving spatial analysis and orientation (McFie, Piercy, and Zangwill, 1950). This appears to occur as a relatively clear-cut defect unrelated to any generalized impairment of language, intelligence, or memory.

The object of this paper is to report a case of right-sided cerebral thrombosis followed by left hemiplegia with extensive sensory loss and anosognosia, and accompanied by profound impairment of visual-spatial perception and performance. It was possible to follow the gradual resolution of these abnormalities in some detail, and as there was little, if any, generalized confusion at any stage of the illness, it is felt that certain features emerged of sufficient interest to justify the publication of a single case.

The Onset of the Illness

Mrs. H., a married woman of 62, had been well until December 23, 1950. She was known to have had mild hypertension for 10 years, with a diastolic pressure in the region of 100 mm. Hg. Her pre-morbid intelligence appears to have been high ; the wife of a scientist with three grown-up children, she was active, well-read, and interested in her home and the careers of her children. There was no history of any previous cerebral vascular episode. She was right-handed. The following account of the onset of her illness was obtained from her husband.

At 5.30 p.m. on December 23, 1950, Mrs. H. was in the kitchen of her house when she was seen to sway on her feet. She first complained of pins and needles in the left arm, and then that the left leg felt weak, but she was able to walk into another room and sit down. One hour later she was seen by a doctor, who noted some indistinctness of speech and drooping of the left corner of the mouth. The eyes tended to deviate to the right, but the left grip was still " quite good ". There was definite weakness of the left leg, and when she insisted on kneeling down to straighten the carpet she was unable to get up without assistance. By 7.30 p.m., when she was helped to bed, the left leg and arm were much weaker, but the patient remained conscious and rational. Her speech had become more indistinct with dribbling of saliva from the left corner of the mouth, and numbness of the left side was first noticed at this time. By the following morning there was profound weakness of the left face, arm, and leg, with loss of feeling, and she complained of severe right frontal headache which persisted for three days. There was no loss of consciousness, and no confusion was noted by her relatives, but little movement of the left arm or leg was seen over the next few days, and she was admitted to the Middlesex Hospital under the care of Dr. D. McAlpine on December 30 with a provisional diagnosis of cerebral thrombosis.

On admission the patient gave an account of her illness which was substantially correct, but there was some confusion as to time and date. She described the onset in the kitchen correctly, and remembered that her daughter had sent her into another room, but whereas she was aware she had lost power in the left leg, she insisted that the left arm had not been affected and that it was normal. Routine questions about her general health, past and family history were answered correctly ; for example, she knew her name, age, home address, and telephone number, and she talked with normal interest of her family and their doings, in particular being con-

cerned over their reactions to her illness. She also spoke sensibly of her hypertension and of the phenobarbitone which had been prescribed for her in the past.

On general examination signs of mild hypertension were found (B.P. 185/95) but no other abnormality outside the central nervous system. A detailed neurological examination was carried out on December 31, and the findings were as follows :

Speech.—There was no dysphasia. A slight dysarthria was attributed to a weakness of the left side of the face and left half of the tongue.

Mood.—The patient was aware that she was seriously ill and was depressed, but her mood was abnormally labile, and she could be made to laugh or cry easily.

Cooperation.—She cooperated well during examination, but in tests of memory and intelligence she tended to answer rather quickly and would guess rather than think again if she was wrong ; this was particularly noticeable when she was tired.

Confusion.—She knew that she was in the Middlesex Hospital and that she had arrived the day before, but was still slightly confused in time. There was difficulty in assessing the number of days that had elapsed since the start of her illness, which she considered to have been on Christmas Day. Otherwise, she showed an almost complete absence of confusion, save in her attitude to her hemiplegia.

Calculation and Retention.—Simple tests with numbers showed that she had a disproportionate difficulty in retaining digits and in adding and subtracting. She could repeat four figures forwards but not backwards. She added 7s serially to 8 correctly as far as 36, but then started adding 9s, and she was unable to subtract 7s serially from 100 at all. The Babcock sentence was repeated to her twice, after which her version was : " Thing a nation needs to be rich is a large supply of wood—and food ".

Cranial Nerves.—A left homonymous hemianopia was present extending to the fixation point, but acuity for central vision was not grossly reduced. There was no failure of conjugate eye movement to the left but it was poorly sustained. Coarse horizontal nystagmus was present on conjugate deviation to the right. There was complete superficial sensory loss over the left side of the face with absence of the left corneal reflex. Motor power was slightly reduced in the upper facial muscles on the left, with gross lower left facial weakness, and protrusion of the tongue to the left. The head was held slightly turned to the right at rest, and there was some loss of power of rotation to the left. Auditory acuity was normal in both ears. Auditory inattention was not present ; when tested by two examiners speaking on either side of her, she repeated the first figure of a double number spoken into the left ear, and the second figure of a different double number spoken simultaneously into the right ear, thus forming a composite number.

Trunk and Limbs.—Tone and power in the right arm and leg were normal. Voluntary power was absent in the left hand and in all the muscles of the left arm and shoulder, and no contraction of the left trapezius could be obtained. Associated movements were not observed. Tone was slightly increased in the left biceps, the arm being held in partial flexion, but the wrist was still flaccid and deep reflexes were only moderately increased compared with the right.

In the lower limb slight movements of flexion and extension were possible at the left hip, but no other voluntary movement was observed in the leg or foot. Tone was already markedly increased, more at the ankle than at the knee, with correspondingly increased deep reflexes and sustained clonus at the left ankle. A well-marked extensor plantar response was present with flexion of the left knee and hip. The right plantar response was flexor.

There was no appreciation of touch, heat, cold, scratching, or repeated pinpricks over the left side of the neck and trunk to the mid-line, nor over the left arm and leg. Some generalized response to strong painful stimuli was elicited by pinching skin, muscle, or tendon, but did not occur in response to pinprick. (This phenomenon had certain features of interest, and is further described below.) Vibration sensation was lost over the limbs and abdominal wall on the left, and passive movements of the left shoulder and hip were not appreciated. Sensation on the right side remained normal, and sphincter control was unaffected.

Subsequent Progress of Left Hemiplegia

The recovery of motor power was slow and incomplete. Four weeks after admission slight weakness of the left upper facial muscles was still present, with moderate left lower facial weakness, and the tongue was still protruded to the left. Some power had returned in the left trapezius, but no movement at the shoulder joint had been seen. A trace of flexion and extension of the elbow was present but no voluntary movement of the wrist or hand. Tone was markedly increased throughout the arm particularly in the flexors of the elbow, wrist, and fingers, with clonus of the brachio-radialis. There had been slight improvement in power at the left hip, but no voluntary movement at the left knee or ankle. Gross extensor hypertonus was present in the left leg.

By the tenth week the patient had begun to stand, and to walk a few steps supported on each side. Loss of power was complete at the shoulder joint and not less than 80% at the left elbow, with some flexion but no extension of the wrist and fingers. The strength of the left hip muscles had further improved and was about 50% normal. Some flexion and extension were possible at the left knee, with a trace of voluntary dorsiflexion at the ankle.

By the twentieth week there had been further improvement in power. There was still no voluntary movement at the left shoulder joint ; flexion at the elbow was possible against gravity, but extension only with gravity eliminated. Flexion and extension at the wrist were strong enough to overcome gravity, but no fine movements of the hand were possible. Tone remained grossly increased, particularly in the flexor groups. There had

been little change in the leg save for the reappearance of a trace of flexion of the toes. Loss of power was estimated as 50% in the hip muscles and in the quadriceps, and 70% in the knee flexors. There was gross extensor hypertonus with contracture of the calf muscles.

When she was discharged on June 2, 1951, the patient could walk with a hemiplegic gait using a right elbow crutch and with one person supporting her on the left side.

Sensory Disturbances and Anosognosia

Agnosia and Anosognosia.—On admission the patient's attitude to her left side was grossly abnormal: when lying in bed she appeared completely unaware of it, and it frequently became uncovered with the leg half out of bed without any attempt being made to replace it or to pull the bedclothes over it. During examination the patient never looked at the left arm or leg, even when asked to concentrate on some movement or during sensory testing. The neglect of her left side obtained even when her head was turned to the left.

On direct questioning she denied that there was anything wrong with the left arm. She admitted to some weakness of the left leg but responded to requests to move it by making the corresponding movements on the right, as with the arm, and in each case would appear satisfied with the result. When the immobile left arm was shown to her, she recognized it as her own, but rationalized her failure to move it, as is shown by her remark, " Can only move it if there is object to move ".

Right-left Confusion.—On admission there was difficulty in identifying promptly her own and the examiner's right and left hands. Her performance in this respect was very variable, but even some weeks after admission she was only able to select the examiner's right hand by making a gesture of shaking hands.

Pain Perception.—Initially the patient showed a curious generalized response to strong painful stimuli to the left side. This was only briefly mentioned in the preceding section. It consisted of general restlessness and uneasiness for which the patient could find no cause, and which she never associated with a peripheral stimulus even in repeated tests. The restlessness was accompanied by tachycardia, a rise in blood pressure, dilatation of the pupils, and deepening of respiration, all appearing within one or two seconds of a strong pinch of skin or deeper structures. The response could not at first be elicited by pin-prick. It could be produced from the face, neck, trunk, and limbs on the left side. In the case of the left leg it was accompanied by reflex flexion-withdrawal of the limb, and usually by flexion of the right leg as well.

It was noticed that in addition to the restlessness and movement in bed the patient would sometimes start scratching the chin, neck, or left flank with her right hand. At first this was not an invariable accompaniment of the general response, but later it became more prominent.

Asked if she felt anything, the patient gave replies such as, " Am moving to make me more comfortable ", or, when scratching her face, " Chin felt queer ". On another occasion she moved restlessly in bed and sighed, and when asked the reason replied: " Not very happy ", adding, " Would you be in my position? " She was quite unaware that her left side was being stimulated and denied pain on every occasion.

Subsequent Progress of Sensory Loss and Anosognosia

On January 2, 1951, the patient was still mildly confused in time, saying that the day of the week was Monday instead of Tuesday, and although she was corrected was wrong again at the end of the examination. She still maintained that the left arm and leg were " all right " and moved the right instead. Scratching was a prominent part of her response to painful stimuli. Pinching the left ear resulted in scratching of the chin, pinching the arm in scratching of the left side of the neck, and pinching the left leg or foot produced scratching of the left flank. This association was not invariable, and if asked whether she felt anything she would reply that she was " itchy " or " twingey " all over, or rationalize by saying that her flank was sore where she had been lifted and turned in bed. On all occasions she showed general restlessness and uneasiness but denied pain anywhere.

On January 3, when asked about her face, she stated that the left side felt numb " like someone else's face ". She said that the left hand was " all right ", but on being asked to show it, showed the right instead. When shown the left hand she recognized it, and after trying to move it said, " Will not lift ", and that it was " very naughty ". She described the left leg as " all right " but moved the right instead. Painful stimuli to the left side produced restlessness and scratching, as before, without any awareness of the peripheral stimulus. On this occasion there was no general confusion; she named the time and date correctly and remembered the names of her visitors of the evening before.

On January 4 the patient again denied her hemiplegia. On being told to move the left hand, she merely wagged it with the right hand and appeared satisfied with the result; when the hands were separated and another attempt made, she moved the right hand as on previous days. Pin-prick was occasionally felt as dull touch over the upper face, but there was no other change in sensation.

On January 5 the patient's attitude to her hemiplegia was changing but was still grossly abnormal. She tended to make light of her disability, and the following conversation took place:

" How is the left hand? " " Better."
" Try to move it." (No result.)
" Is it moving? " " It will do in a minute."
" What is the matter with it? " " Dopey."
" How is the left leg? " " Much better."
" Is it dopey? " " No, I am moving it all the time " (moving right).

Examination also revealed some improvement in sensation; pin-prick was felt as dull touch over the left face, neck, and arm, but whereas it was correctly lateralized and fairly well localized over the face, in the

arm it was referred to the right side instead of the left in about half of the replies. This was the first occasion on which allaesthesia was observed. Pin-prick was not perceived as such over the left foot, but produced some scratching of the left neck and clavicle, which the patient described as " twingey ". Vigorous pinching of the left big toe produced general restlessness, but on one occasion the patient recognized and localized the stimulus correctly, pointing out the toe with the right hand.

Her lack of general intellectual impairment was shown by a clear and correct account of the nature and scope of her son's work, which she was able to give on this day.

On January 6 further improvement was noted. Asked if she could move the left hand, she said " Yes ", but when it was attempted there was no movement. The patient said she did not know why she could not move it, and referred to it as " silly hand ". There were some failures to name her own and the examiner's right and left hands correctly and she said, " I confused my right and left, I can't make it out. I'm silly ". To avoid verbal confusion between right and left during sensory testing, the patient was asked to try to point out the site of the stimulus on each occasion. Pin-pricks on the left face were correctly lateralized and were localized to the forehead, cheek, or chin, but were felt as dull touches. Pricking of the arm caused only scratching of the left clavicle. From the left knee a proportion of pin-pricks was felt as dull touches on the left calf, but an equal number was referred to the right knee, or caused withdrawal movements without localized sensation. Hard pinching of the left big toe was similarly either localized, referred to the right toe, or ignored in spite of reflex withdrawal and general body movement.

On January 8 and January 9 the patient still found difficulty in naming the day of the week and the time, and named the examiner's right and left hands wrongly in three consecutive tests. Her attitude to her hemiplegia was one of unconcern, as shown by the following conversation :

" How is the left arm ? " " Better."
" Is there anything wrong with it ? " " No, I don't think so."
" How is the left leg ? " " Better. Ask Nurse " (referring to the physiotherapist).
" Is there anything wrong with it ? " " Don't think so."
" Could you walk on it ? " " Let me try."

Pin-prick was usually felt as dull touch and localized to within two to three inches over the face, neck, and flank. Sensory inattention (Critchley, 1949) was present to simultaneous bila.eral pin-pricks of approximately equal strength. From the left fore-arm stimuli were referred to the hand, and from the upper arm to the clavicular region. Replies were rather more variable from the leg. Stimuli over the left face and trunk were occasionally referred to the mid-line, but no allaesthesia remained save from a small area on the anterior abdominal wall. Even here it was inconstant, and it was not observed again in subsequent examinations.

The absence of general intellectual impairment may be illustrated by a conversation at this time in which she discussed various books that she had been reading just before her illness (in particular Stefan Zweig's *Beware of Pity* and a life of Balzac), and bewailed the fact that she had not been able to read since the onset.

By January 16 the disability of the left arm was no longer ignored but was treated in an objective fashion ; the patient referred to it as " poor little withered hand ", picked it up, and kissed it. There was still some neglect of the left leg both in bed and when sitting out in the ward. At this time pin-prick was poorly localized and felt as tapping on the left arm and leg, but both pin-prick and pinching could still sometimes produce general restlessness. In the case of the arm this was usually accompanied by a " twingey " feeling in the region of the clavicle, and by scratching, but from the leg it could still occasionally be elicited without any localized sensation whatever. This response was not observed again after this date.

On February 13 she still showed considerable emotional lability, becoming very upset over other patients in the ward, and laughing and crying readily. There was no denial or obvious neglect of her hemiplegia, and the patient stated that if she wanted anything from the locker on the left of her bed, she would try to use the left hand without thinking. Pin-prick was felt as touch and correctly localized over the face. It was referred from the left upper arm to the shoulder and interpreted as painful, but was not felt at all on the forearm. On the legs pin-pricks were poorly localized but were painful. Sensory inattention was present. Light touch was not perceived at all over the left side, and postural sensation was absent.

At this time it became apparent that a phantom limb was present. During the early weeks of her illness, on being requested to find the left hand with the eyes closed, the patient had been accustomed to feel for the left shoulder and work her way down the arm to the hand. After the disappearance of anosognosia, if the left arm were held up in the air with the patient's eyes closed, she would feel for the left hand on the lower abdomen " in the usual place " for a few seconds before tracing it by way of her left shoulder. If the left arm were held up in the air and she were asked to place the right arm in the same position as her left arm, she would place it on the abdomen, with fingers separated and extended. She was unable to move the phantom, and readily ignored it when she could see the real position of the left arm. The phantom persisted until her discharge from hospital. The appearance of a phantom with improvement in cerebral function is in interesting contrast to the patient described by Head and Holmes (Head, 1920) in whom a stroke abolished a phantom leg.

On March 18 a considerable degree of sensory loss was still present, but the patient's attitude to her left side was not obviously abnormal, and there was no neglect nor denial of disability. Pin-prick was felt dully over the left face with inattention. There was some blunting over the left arm and forearm, and a pin-prick felt dull on the hand ; on the leg it was recognized as painful or " squeezing ". It was localized to within three or four inches on both arm and leg. Pinching of the skin was not distinguished from pin-pricks, but was

recognized as painful. No general restlessness was elicited by either of these stimuli. Postural sensation was absent at the left shoulder but present at the hip. Light touch remained wholly absent over the left face and limbs, and hot and cold stimuli were not appreciated on that side.

On May 16 appreciation of light touch was still absent, but firm pressure was felt and was well localized except on the leg. A prick was recognized by its unpleasant quality and was well localized on both arm and leg. There had been no further improvement in position sense, nor in appreciation of heat and cold.

At a follow-up examination on March 22, 1952, the patient's phantom limb was still present. The phantom hand was felt to be resting on the abdomen with the fingers fully extended, in contrast to the flexed posture of the real fingers. In addition the patient described frequent sensations of individual finger movement although she knew by inspection that these never in fact occurred.

Visual Field Defects

On admission a left homonymous hemianopia extending to the fixation point was present on confrontation; the patient denied this even when it was demonstrated to her. One week after admission she was able to appreciate moving objects in the upper part of the left half-fields, with visual inattention, and three days later stationary objects were perceived in the left upper quadrants. Three weeks after admission movements were consistently appreciated in the left lower quadrants but stationary objects were not seen. After one month it was noted that the patient showed no disorientation in the left half-fields, and was able to touch on request the nearer or farther of two objects introduced into them. By the time she was discharged, the patient was able to recognize common objects in the upper but not the lower quadrants, and a partial left lower quadrantic defect was present on perimetry ($\frac{1}{2}$° white). Visual inattention was still present.

Unilateral Neglect of External Space

Drawing.—Shortly after admission a drawing of a clock showed an unmistakable neglect of the left half, all the numbers on the left side being drawn in their mirror-image positions on the right. On January 6 further clock drawings produced an identical result; two days later, however, the patient was able to draw a clock normally, numbering the dial correctly but had great difficulty in drawing in the hands to show a given time.

Reading.—On admission she was able to write her name and address, but her handwriting was almost illegible. She herself had great difficulty in reading. The words "The Middlesex Hospital", printed in capitals at the top of ordinary ward paper, were first read as "hospital", then "Essex hospital", and finally, with encouragement, "The Middlesex Hospital".

On January 6 the patient was tested with the following sentence printed in half-inch block capitals:

GEORGE LIKES
EATING HIS
SUNDAY DINNER

This was first read as "likes his dinner", but on being made to search for the rest of the words, she managed the complete sentence, only to come back to it a few minutes later and read "George likes his dinner", denying the presence of any other words on the page even when it was suggested that she had omitted some.

On January 9, on testing with Snellen's type at six metres, the patient read only the right-hand letter of each of the first five lines. Three days later she read Snellen's type normally (V.A.R. = 6/24, V.A.L. = 6/18), but still had great difficulty in reading sentences. She was unable to understand newspapers or letters, as, in attempting to read, she would jump from the end of one line to the middle of the next, and would misread or omit words, syllables, and individual letters, so that even simple sentences became nonsense.

On January 17 it was discovered that her performance was strikingly different when reading from above downwards with the letters in columns starting from the left of the page. The first test sentence is below:

(A)	AN INTELLIGENT	A	S	S
	APPROACH TO THE STUDY	N	T	O
	OF THE NERVOUS SYSTEM		U	M
	REQUIRES SOME	I	D	E
	KNOWLEDGE OF ITS	N	Y	
	ANATOMY	T		K
		E	O	N
		L	F	O
		L		W
		I	T	L
		G	H	E
		E	E	D
		N		G
		T	N	E
			E	
	(B)	A	R	O
		P	V	F
		P	O	
		R	U	I
		O	S	T
		A		S
		C	S	
		H	Y	A
			S	N
		T	T	A
		O	E	T
			M	O
		T		M
		H	R	Y
		E	E	
			Q	
			U	
			I	
			R	
			E	
			S	

The patient was told to spell out the letters making words where possible. Reading horizontally (A) her version was: "In itellgent enouch to the stry of the blous sytem reuies some boese of its anatmy" (time taken 2 min. 45 sec.). In contrast to this the first time she read the sentence from above downwards (B) she made only two small mistakes: "Hn intelligemn approach to the study of the nervous system requires some knowledge of its anatomy" (time taken 3 min. 30 sec.). This phenomenon was not analysed further, for by January 20 her difficulty in reading horizontally had disappeared and did not recur.

Tactile and Visual Neglect in External Space.—On admission a simple test described by Sendifer (1946) was tried: a number of common objects was spread across the coverlet occupying the whole width of the bed, and the patient was asked to find as many of them as possible by groping with the right hand with the eyes shut. She searched approximately equally to her right and left, and recovered all the objects save one on the extreme left at the limit of her reach. In a further test one week later she recovered all the objects without an omission.

On January 11 an attempt was made to investigate the discrepancy between her visual and tactile performance in the left half of space. The patient was presented with a large board covered with paper in which a number of drawing pins was placed at random. The patient was then asked to ring each pin with the right hand, using different coloured pencils with the eyes open and shut respectively. With the eyes open she started in the middle of the board and ringed only those pins to the right, saying at the end, "That's the lot", and denying that she had omitted any. With the eyes shut she missed occasional pins indiscriminately, but worked gradually over to the extreme left of the board, and finished by running her fingers down the left-hand edge. In a subsequent test she was restarted on the extreme left with her eyes open and told to ring those that she had missed, but even with this encouragement she only found a few pins before drifting back to the centre of the board and ignoring those remaining to her left. It was not until February 1 that this test was performed normally with the eyes open.

Visual-spatial Agnosia

The patient showed disorders of visual-spatial perception of the type analysed by McFie, Piercy, and Zangwill (1950) and referred to by them as visual-spatial agnosia. Initially there was no difficulty in revisualizing people or places nor in recognizing common objects, and distances were judged approximately correctly. She made no errors in testing for deformation of coordinates, nor in bisecting lines. It was not possible to demonstrate any gross defect of route finding in her new surroundings. Topographical memory was not impaired, and she was able to describe her route home from the hospital with detail and accuracy. Similarly, she could describe the main turnings off Piccadilly Circus correctly, and mention a number of well-known buildings and shops in the vicinity. In contrast to this she was incapable of the simplest plan drawing. In drawing Piccadilly Circus, for example, she would start with Oxford Street drawn vertically down the paper, and add everything else in a haphazard jumble to the right of it. Her copying of simple geometrical patterns was very faulty, and drawings of objects from memory showed marked dysjunction.

Three weeks after admission, on the Progressive Matrices (1938) Test, she chose only eight correct patterns out of the 12 in Set A. Six weeks after admission, when tested with Kohs Blocks (Wechsler Form 2) she was unable to proceed beyond the demonstration pattern, even when shown the solution to pattern 1. When re-tested on progressive matrices just before her discharge from hospital, she showed little improvement, with only 14 correct choices out of the 24 in Sets A and B, whereas a vocabulary test (Wechsler) gave an intelligence quotient of 119.

Owing to her hemiplegia it was not possible to perform any satisfactory test of apraxia for dressing.

The Relation of Unilateral Neglect of External Space to Visual-spatial Agnosia

Attempts were made to assess the extent to which the patient's neglect of the left half of visual space was contributing to her general difficulty in dealing with visual-spatial problems. Two tests in particular were used and merit brief description.

On January 8 it was noted that she was able to draw a clock normally, but had great difficulty in drawing in the hands. When this was investigated further with a large cardboard clock, it did not appear to be related to a neglect of the left half of the clock, as her performance was no better when setting the hands or telling the time on the right half of the clock face. She appeared to realize her disproportionate difficulty, saying on one occasion, "Fancy not being able to do simple stuff like this", and adding that she did not seem to be able to think what it should look like. The patient had equal difficulty in solving clock problems in her head, and would be defeated by a question such as, "If the hour hand is at 3, and the minute hand at 10, what is the time?" This was not thought to be part of a dyscalculia, as she was able to add and subtract 5s serially to and from 60 with ease. Her performance in clock problems improved only slowly, and mistakes were still made in decreasing numbers until the third month of her stay in hospital.

A second test, suggested by Dr. Michael Kremer, was devised with a simple circular maze 18 in. in diameter, the turnings being so arranged that in finding a way from the centre to the outside, there was a short way involving left turns and a long way using right turns (Fig. 1). It was hoped that this might reveal a defect in route-finding due to neglect of the left turnings, and by making the maze of thick circular strips of orthopaedic felt stuck on a flat board it was possible to compare the patient's performance using sight and touch. This maze was first used on January 15, when tests with drawing pins and in reading still indicated some visual

FIG. 1

neglect of the left, but it showed in fact not a neglect of turnings to the left, but a complete inability to select turnings leading outwards rather than those leading back to the centre. Although the nature of the problem was well understood, the patient always took random turnings, and would sometimes return to the centre as many as three times before reaching the outside. By touch she was only slightly better than with the eyes open, and with both she appeared to be incapable of pausing to consider any one turning in relation to the whole problem. Her performance improved only slowly, and in spite of repeated use of the same maze it was still abnormal two months later.

Discussion

On clinical grounds there seems to be little doubt that this patient suffered a cerebral thrombosis resulting in damage to a large area of the right cerebral hemisphere. Consideration of a more exact anatomical localization is not profitable in the absence of necropsy material. An electroencephalogram taken shortly after admission was indicative of a right-sided lesion. It showed frequent replacement of the alpha rhythm in the right occipital area by low voltage 7/sec. waves with high voltage 2–3/sec. waves in the right parietal and frontal areas. Seven weeks later 1–2/sec. waves were still present in the posterior part of the right temporal region and in the right occipital region ; further follow-up E.E.G.s showed the alpha rhythm returning to those areas in which it had previously been suppressed.

The anosognosia showed certain points of interest. The denial of disability was focal, including the hemianopia and left arm, but not extending fully to the left leg. It was not associated with any clouding of consciousness nor with generalized confusion. The patient's acceptance of her disability was initially qualified by attempts to rationalize her failure to move, and was later characterized by a curious objective attitude, as if her hand were leading a perverse, independent existence for which she was not directly responsible.

Against the background of profound unilateral loss of pain perception, the patient showed restlessness and uneasiness, tachycardia, rise in blood pressure, pupillary dilatation, and deepening of respiration in response to noxious stimuli on the left side, a phenomenon which slowly vanished in rough correspondence to the anosognosia. Four stages in this process could be discerned. Initially general restlessness and uneasiness were unaccompanied by any recognition of the stimulus. Later the general response was accompanied by scratching of the left side, usually the chin, neck or flank, in response to stimulation of the ear, arm, or leg respectively, and later still the peripheral stimulus began to be fairly consistently interpreted as itching or twinges in these areas. Finally the patient began to recognize and localize the stimulus in a large proportion of tests, the generalized response still appearing occasionally without local sign. This phenomenon has received scant attention in the literature of anosognosia. A similar instance has been described by Pötzl (1924, Case 2). Generalized movements with denial of pain have also been noted by Nathan (personal communication) in patients with anosognosia receiving injections of penicillin on the affected side. Our case differs in certain particulars from Sandifer's (1946) description of a patient in whom noxious stimuli " evoked every expression of pain but denial that she felt any ". Allaesthesia in our patient seemed to be a transient phenomenon which appeared at an early stage of sensory recovery and lasted only a few days.

The patient's initial dyslexia may be ascribed to a massive neglect of the left half of visual space, but the discrepancy between her performance in horizontal and vertical reading is more difficult to analyze. It is likely that vertical reading was accomplished by identification of single letters and subsequent synthesis into words, this latter being performed correctly but slowly, indicating some defect in conceptual thinking where visual symbols were involved. The reason for her horizontal dyslexia remains obscure. It may have been due to a partial neglect of the left half, although by this time she was able to find correctly the beginning of each new line of print. Gross oculomotor defects were by this time absent, movements being full in

all directions and unaccompanied by nystagmus. Neglect of the left side may be sufficient to upset saccadic eye movements in horizontal reading more than in vertical reading, and may have been responsible for her poor performance (Zangwill, personal communication). On the hypothesis that letters in the right half of the visual field might be causing visual inattention to the left half, an attempt was made to cause inattention during vertical reading by interpolated visual stimuli in the right half-fields; no deterioration in performance resulted from this manoeuvre, but this negative result may not be significant.

In general, visual-spatial disturbances outlasted the body image disturbances, but this may merely indicate that the cerebral damage was most severe in the posterior part of the right hemisphere. The disorders of visual-spatial perception conformed to those described by McFie and others (1950). The intactness of topographical orientation and memory in this patient is in accord with the findings of Hécaen, Ajuriaguerra, and Massonnet (1951) in six patients with parieto-occipital lesions of the minor hemisphere, and contrasts with those of McFie and others (1950), who recorded such defects in six of their eight patients.

Neglect of the left half of external space was shown on the motor side in drawing numbers on a clock face, and on the sensory side in reading tests. However, when the patient was required to ring drawing-pins on a board with the eyes open and closed, her performance was strikingly better with the eyes closed. It thus appears that in this patient a neglect of the left half of space was determined by visual experience, and that external space (Brain, 1941) may be further divisible into " visual space " and " tactile space ".

Two further problems remained largely unsolved by our analysis, the first of these being the extent to which unilateral neglect of visual space contributed to visual-spatial agnosia. All that can be inferred from our patient is that gross neglect of the left improved relatively early, with persistence of visual-spatial agnosia. However, the continued tendency to put in towns on the right side of a map when map-drawing (which was still evident on February 15) indicated some persistent neglect of the left although grosser tests failed to reveal it. The extent to which conceptual processes involving visual symbols were affected in our patient also proved difficult to assess. Her failures to do clock problems in her head, and to do the maze test with the eyes shut were similar to her performance when doing these tests by sight. Therefore, in spite of her denial of difficulty in revisualizing people or places, it is hard to resist the inference that a conceptual defect was present where visual problems were involved. It would be of some interest to know whether her visual concepts lacked a left half, and if so, whether this was an inattention which could have been modified by extraneous visual stimuli.

Summary

A right-sided cerebral thrombosis in a woman aged 62 gave rise to a left-sided hemiplegia and hemianopia, with profound left-sided sensory loss.

An initial anosognosia was followed by a detached attitude to her disability.

During the stage of profound left-sided sensory loss noxious stimuli to the left side gave rise to restlessness and uneasiness, with tachycardia, rise in blood pressure, pupillary dilatation, and deepening of respiration but without any conscious recognition of the stimulus. The stages in the resolution of the phenomenon were followed.

There was a neglect of the left half of space which was demonstrable only when the eyes were open.

During the stage of recovery vertical reading was performed better than horizontal reading.

Visual-spatial agnosia was a marked feature of the illness, and outlasted neglect of the left half of space.

We wish to thank Dr. Douglas McAlpine for his help and for permission to publish this case, and Dr. Michael Kremer for his interest and advice on the problems presented by the patient.

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SOME MECHANISMS OF TACTILE LOCALIZATION REVEALED BY A STUDY OF LEUCOTOMIZED PATIENTS

BY

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This investigation was undertaken following a suggestion that finger agnosia occurred after prefrontal leucotomy. A preliminary investigation did not reveal the presence of finger agnosia as described by Gerstmann (1924) but confirmed the existence of a post-operative difficulty in localizing tactile stimuli on the fingers. Accordingly, the present investigation was designed with the object of examining the extent and nature of this defect and in the hope that the nature of the errors would reveal something of the mechanism by which normal localization of tactile stimuli occurs.

Although tests of tactile localization are used clinically, and may assist in the diagnosis of cerebral damage, very little is known concerning the neurological and psychological mechanisms mediating the " local sign " of tactile stimuli. The most notable clinical studies are those of Head (1920) who convincingly demonstrated the impairment of tactile localization which may attend parietal lobe dysfunction. Head also studied the restitution of sensory function following section of the radial nerve. His studies of peripheral nerve lesions led him to postulate the famous protopathic-epicritic dichotomy whereby it was claimed that only epicritic nerve fibres mediated local sign, thus reverting in essence to the " doctrine of specific nerve energies ". This suggestion has been seriously challenged by Walshe (1942) on theoretical grounds, and Sherrington (1947) has suggested that the essential peripheral basis of tactile local sign might be the distribution of overlapping sensory units. In the case of defects of tactile localization resulting from cerebral injury Head asserted that such defects occurred only as a result of parietal injuries and that, when errors of localization on the fingers occurred, their distribution was consistent with random guessing and provided no evidence for supposing either that errors were more frequent in the case of some positions than others, or that stimuli were erroneously referred more frequently to one site than to another. He refers to a conceptual " schema " of tactile (and proprioceptive) orientation which presumably is either intact or, when impaired by brain damage, impaired in an essentially non-specific way.

Gerstmann has claimed that with some cerebral lesions there is a condition described as finger agnosia in which there is an inability to recognize individual fingers. It is not clear, however, that in these cases there is an impairment of the ability to make a localizing response to stimuli on the affected fingers.

Investigations in this field undertaken by psychologists have been almost exclusively concerned with healthy subjects, and have been devoted largely to testing the relative efficiency of different skin areas and to demonstrating the presence of systematic errors of tactile localization. Thus it has been known for many years that the accuracy of tactile localization depends greatly on the part of the body stimulated. It is also well known that stimuli tend to be displaced towards the nearest joint rather than away from it, and towards the pre- and post-axial borders of the limb. In one of the more important contributions to the subject Spearman (1906) demonstrated many of the factors involved. He showed that when a point on a limb was stimulated the subject tended to refer it in a direction which was dependent on the relative position of the stimulus site to the rest of the body, and, in particular, that the direction in which an error was made was related to the direction in which the stimulated part deviated from its mid-position with reference to the next proximal joint. Some of these results have been confirmed by Hulin (1935), who also showed that when a limb was stimulated in motion then the direction of movement correlated with the direction of the error. He also showed that the position of the model on which the localization is identified by the subject is also of importance. None of these studies has been primarily concerned

with the mechanisms which enable higher level responses to be made in a way which is dependent on the locus of the tactile stimulus. Many of them have presupposed a fixed " sensory map " of a type which is incompatible with modern views of cortical physiology.

Methods

The method used here for the examination of tactile localization on the fingers is that of Henri (Head, 1920), in which the subject indicates on a three-dimensional model the position corresponding to the site of stimulation. As in Head's work the model used was the corresponding hand of one of the examiners. The subject was stimulated by a second examiner who used a wooden stylus with a 1 mm. rounded end. The intensity of the stimulus was not precisely controlled but the stimulus may fairly be described as a light touch of about $\frac{1}{2}$-second duration. Stimuli which were not perceived at all were always repeated but not stimuli whose mere location was inadequately perceived. Most testing was done with the patient in bed. The stimulated hand lay comfortably on a board in the prone position with the fingers slightly spread. During stimulation the subject's hand and the experimenter's arm were shielded from the subject's view, but the model hand was continuously before the subject who was required to indicate the site of stimulation by pointing to the model with a pencil held in the free hand. The subject's response was recorded diagrammatically by one of the examiners.

During each test session the four fingers (but not the thumb) of both hands were stimulated four times. These four stimuli on each finger were placed two on either side of the finger, and the two stimuli on each side of the finger were placed one in the middle of the distal phalanx and one in the middle of the second phalanx. Each position was stimulated once ; a total of 32 stimuli for every test session. Differential fatigue and practice effects on the two hands were obviated by alternating the stimulation of the two hands in groups of four stimuli. The order of stimulation of the two hands is shown by the series ABABBABA where each letter represents four stimuli to one hand, and a change of letter a change in the hand stimulated. In each hand, and on each occasion of testing, the order in which the sites were stimulated was random. As a further precaution the hand first stimulated on each occasion of testing was alternate to the hand first stimulated on the immediately previous test session. The thumb was not stimulated, because during the pilot stage of the investigation it was found that stimuli on the thumb were always correctly referred even when considerable difficulty was experienced in localizing stimuli on the fingers. Subjects were instructed that the stimuli would always occur on the sides of the fingers and either in the middle of the second or third phalanx and they were required to make their responses in accordance with these standards.

In estimating error in tactile localization two principal methods may be used. Either the direction and distance of the error may be recorded or else responses may be scored according to arbitrary standards of accuracy. Since it is not justifiable to regard the hand as an equipotential sensory surface, and since there are obvious difficulties attached to measuring extent of error when, for example, a stimulus is referred to the wrong finger, the second alternative has been adopted. This procedure is further justified by the fact that pilot studies suggested that errors were usually on a conceptual basis, e.g. the subject might refer the stimulus to the wrong finger but to the correct position on the finger.

The errors considered here are of three types : those involving a response referring the stimulus to the wrong finger, those referring the stimulus to the wrong side of the finger, and those referring the stimulus both to the wrong finger and to the wrong side of the finger. Errors involving a response referring the stimulus to the wrong phalanx are not considered here since it became obvious at an early stage that there was a strong tendency to refer the stimulus towards the nearest joint and this effect is already well known.

Twenty-two patients undergoing prefrontal leucotomy were examined before and after the operation.

AGE, SEX, AND DIAGNOSIS OF THE 22 SUBJECTS

Subject	Age at Operation	Sex	Diagnosis
19246	21	F	Facial tics ; depression
30282	38	F	Depersonalization syndrome
18143	41	M	Torsion dystonia ; depression
11564	42	F	Anxiety state with chronic depression
24317	44	M	Tinnitus, depression, following meningitis
28174	45	M	Involutional depression
28286	47	F	Chronic anxiety state
12233	48	F	Schizophrenia
11773	48	M	Chronic anxiety state
19179	50	M	Chronic tuberculosis
15661	50	F	Disseminated sclerosis ; hypochondriasis
8776	50	F	Obsessional neurosis
14183	52	F	Involutional depression
18198	53	M	Involutional depression
18957	53	F	Post-herpetic neuralgia ; depression
8663	54	F	Hypochondriasis
23977	55	M	Post-herpetic neuralgia
14765	56	M	Hypochondriasis ; depression ; ? organic dementia
25598	56	M	Post-herpetic neuralgia
9997	63	M	Involutional depression
13043	63	F	Post-herpetic neuralgia
14481	64	F	Hypochondriasis

It was not possible to record enough errors from most subjects to establish individual pre-operative patterns and the results reported are those obtained for the group as a whole. In order to compare the results obtained before and after leucotomy, and to allow for differences in the number of test sessions of individual subjects, the results have been presented as error rates or errors per 100 stimuli. The percentage error at each stimulus site was calculated for each subject and thence a mean percentage error for the group was obtained. In the presence of individual differences in the distribution of errors such a method would not exclude undue weighting of the general pattern by patients who made frequent errors and might also produce artifactual changes in post-operative error distribution resulting from unequal impairment of subjects with different error distributions. A more accurate mean pattern would be obtained by averaging the pattern obtained by expressing individual error frequencies at the different stimulus sites as percentages of the total number of errors made by the subject. Such an analysis has been made and it is clear that no material distortion in pattern can be attributed to the method of analysis first described.

In order to avoid repetition and to allow comparison between pre- and post-operative performance the former method of analysis is alone presented here. Where necessary raw figures are also reported to allow of the assessment of the significance of the percentage figures. All statistical tests of significance were applied to the raw data.

Results

Errors of tactile localization were made pre-operatively by all our subjects. Post-operatively there was an increase in the frequency of errors. These errors are described first according to the site of occurrence and secondly according to the type of error.

Incidence of Errors.—Pre-operatively a total of 2,064 stimuli were applied to our 22 subjects and of these 273 (13%) were incorrectly localized. This high incidence of errors was largely due to lack of practice on a novel test. After leucotomy 2,118 (21%) of 10,096 stimuli were incorrectly localized, an increase in the frequency of errors which is clearly significant ($p<0{\cdot}001$). Only very occasionally was the patient too confused to cooperate on the first post-operative day. If the subject was able to attempt the task then some stimuli were correctly located. Performance was most seriously impaired immediately post-operatively and improved as convalescence progressed. The error frequency for the first four weeks was 26·1%, 22·5%, 22·0%, and 16·4% respectively.

The frequency with which errors occurred was not the same at each site of stimulation nor for each hand. The distribution of errors over the two hands is shown in Fig. 1. In order that the pre- and post-operative patterns can be compared, and to exclude excessive weighting of the pattern by any one subject, the error at each site is also expressed as the mean percentage error for all 22 subjects.

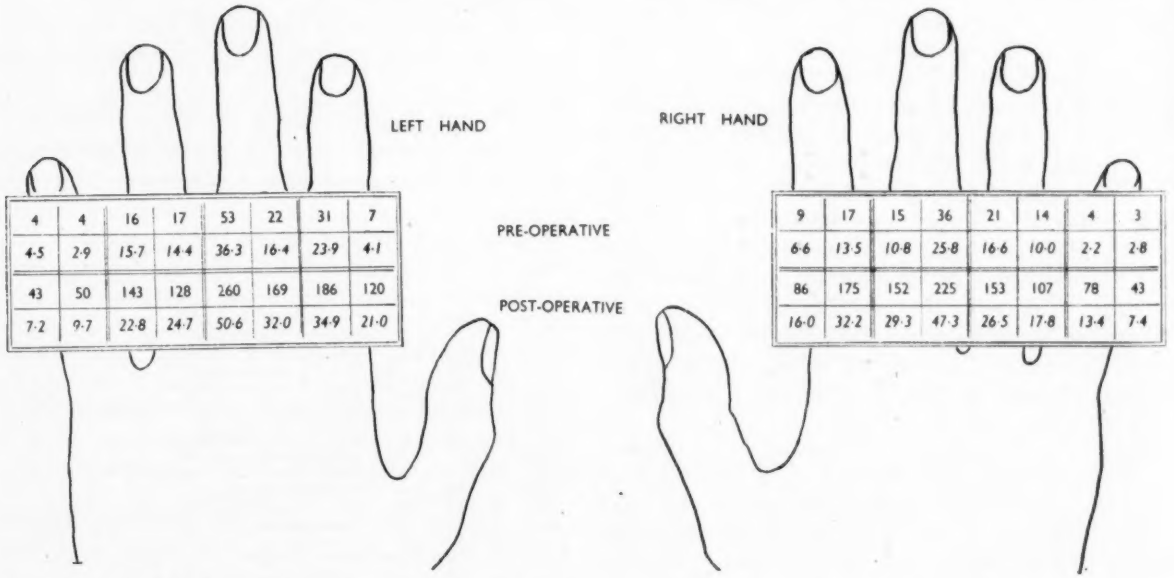

FIG. 1.—This figure shows the number of errors which occurred at each stimulus site in 22 subjects. The figures in italics show the mean error rates for these subjects expressed as a percentage of the number of stimuli at each site.

It is clear that the distribution of errors is not a random one and that mistakes in localizing tactile stimuli are much more frequent at some sites than others. Further it is apparent that these differences between the various sites are very similar on the two hands and that the patterns post-operatively closely resemble those found before leucotomy. Thus on each hand before and after operation the fifth finger is the one at which fewest errors are made. Similarly on both hands both before and after the operation most errors occur following stimulation of the ulnar side of digit 3 where the error frequency is between six and 10 times that at the ulnar side of digit 5. A χ^2 test of significance can be applied to check that the observed distribution of errors over the eight sites on the hand is not likely to have arisen by chance. Pre-operatively, when the errors at corresponding sites on the two hands are summed, the value for χ^2 is found to be 152·3. A similar result is obtained post-operatively when the increase in the number of observations gives an even higher value for χ^2 (451·8) and so an even more significant result.

It is clear therefore that our subjects, most of whom are free from neurological lesions, are liable under the conditions of this investigation to make gross mistakes when localizing tactile stimuli on the hand, i.e. indicate the wrong finger or the wrong side of the finger. This generalization from the summed results is justified by the observation that each of our 22 subjects made at least one error pre-operatively.

The frequency with which errors occur is determined to a considerable extent by the site of stimulation. As, however, the distribution of the errors is related to the type of error made, this effect is reported before making a detailed analysis of the distribution.

Type of Errors.—Table I shows the distribution of errors and the sites to which errors made at each of the 16 stimulus points were referred. As an example, 53 errors or 36·3% of the stimuli applied pre-operatively to the ulnar side of the third finger on the left hand were incorrectly localized. These errors were referred to the other seven sites as shown in row 3U in the top left hand matrix.

It is apparent that both before and after operation errors are not referred to other sites in a random fashion but that when a stimulus is incorrectly localized the site of reference depends on the site of stimulation. This departure from a purely random reference of errors is not due to chance, as is shown by calculating the value for χ^2 for our observed distribution. This has been done by summing the pre- and post-operative results and the results for both hands and calculating χ^2 for the distribution of error referrals in the case of each stimulus site (Table II). The eight values for χ^2 thus obtained are all significant at the 0·001 level of confidence. The pre-operative figures are not examined separately because pre-operatively the number of errors at some sites is too few to permit the application of the χ^2 test.

This relationship might seem to justify the assumption that the error is a quantitative one and that the incorrectly perceived stimuli are most frequently referred to the next nearest site. In this case the error tendency would be suitably represented by the extent of skin surface intervening between the site of stimulation and the site of reference. On this hypothesis there would be an inverse relationship between the extent of the errors and their frequency. Closer inspection of the data presented in Table I shows, however, that in the case of 10 of the 16 sites of stimulation pre-operatively and 11 of the 16 sites post-operatively more errors were referred to a position at two removes from the site of stimulation than to the position immediately adjacent to the stimulus site. That is to say, errors are more frequently referred to the corresponding position on an adjacent finger than to the wrong side of the stimulated finger. Similarly, when errors are referred to an adjacent finger most of these errors are referred by the subject to the side of the finger corresponding to the stimulated side of the stimulated finger. This occurs irrespective of whether that side is nearest to or more remote from the stimulated finger than the alternative side. Thus in both hands pre- and post-operatively three times as many of the stimuli applied to the ulnar side of the third finger are referred to the ulnar side of finger 4 as are referred to the radial side of this finger, although this latter side is nearer to the site stimulated. Similarly, many more of the stimuli applied to the radial side of the fourth finger are referred to the radial side of the third finger than are referred to its ulnar side.

The fact that one aspect of the stimulus situation is correctly perceived even when the total interpretation is wrong suggests that localization is taking place on a "conceptual" basis and that localizing responses are made to more than one spatial attribute or dimension characterizing the site of the stimulus. These different aspects of the stimulus may be perceived with different degrees of accuracy. Two such dimensions appear to be present in the particular performance under investigation. These are (1) the finger stimulated, and (2) the side of the finger stimulated.

TABLE I

SITES OF STIMULATION AND DISTRIBUTION OF ERRORS* PRE-OPERATIVELY AND POST-OPERATIVELY FOR LEFT AND RIGHT HANDS BY 22 SUBJECTS

Pre-operative

Left Hand

SITES OF REFERENCE OF ERRORS

SITES OF STIMULATION	5U	5R	4U	4R	3U	3R	2U	2R	Totals
2R				1 0·6	1 0·6	3 2·3	2 0·7		7 4·1
2U				1 1·1	8 7·0	7 6·5		15 9·2	31 23·9
3R				12 8·3	4 3·6		2 1·4	4 3·0	22 16·4
3U			33 21·8	8 6·4		9 5·5	2 1·4	1 1·1	53 36·3
4R		3 1·7	4 3·2		3 2·7	7 6·8			17 14·4
4U	5 4·5			11 11·2					16 15·7
5R	2 1·4			2 1·5					4 2·9
5U		2 2·3	2 2·3						4 4·5

Right Hand

SITES OF REFERENCE OF ERRORS

SITES OF STIMULATION	Totals	2R	2U	3R	3U	4R	4U	5R	5U
2R	9 6·6		3 2·7	6 4·0					
2U	17 13·5	12 9·6		1 1·1	3 2·0				1 0·8
3R	15 10·8	4 3·4			4 1·2	6 5·9	1 0·3		
3U	36 25·8		1 1·1	8 5·2		2 0·8	24 17·9		1 0·8
4R	21 16·6		1 0·4	9 8·3	2 2·3		6 3·7	3 1·9	
4U	14 10·0			1 0·8	3 3·0	4 2·8		1 0·8	5 2·7
5R	4 2·2								4 2·2
5U	3 2·8							3 2·8	

Post-operative

SITES OF STIMULATION	5U	5R	4U	4R	3U	3R	2U	2R	Totals
2R	3 0·5	12 1·7		16 2·7	3 0·4	38 6·4	48 9·3		120 21·0
2U	1 0·1	11 1·5	18 3·3	16 4·0	49 8·7	23 3·3		68 14·1	186 34·9
3R	1 0·1	10 1·4	9 1·4	71 15·2	31 5·2		14 2·7	33 6·0	169 32·0
3U	8 0·9	8 0·9	118 27·6	42 7·8		71 11·4	12 1·9	1 0·2	260 50·6
4R	4 0·7	25 3·9	20 4·9		14 3·1	54 9·8	3 0·3	8 2·0	128 24·7
4U	14 2·6	20 2·8		75 11·0	26 4·7	6 0·9		2 0·8	143 22·8
5R	12 2·4		6 1·1	27 5·0		2 0·5		3 0·7	50 9·7
5U		25 3·6	16 3·4	2 0·2					43 7·2

Right Hand

SITES OF REFERENCE OF ERRORS

SITES OF STIMULATION	Totals	2R	2U	3R	3U	4R	4U	5R	5U
2R	86 16·0		25 4·4	29 6·5	4 0·6	20 3·4	5 0·7	1 0·2	2 0·3
2U	175 32·2	42 9·2		32 5·3	51 10·1	20 2·8	17 3·2	11 1·4	2 0·3
3R	152 29·3	40 6·9	10 1·5		22 3·9	66 15·1	8 1·2	6 0·7	
3U	225 47·3	2 0·2	12 2·7	47 8·3		36 6·9	123 28·5	3 0·4	2 0·3
4R	153 26·5	5 0·7	2 0·3	70 10·4	11 1·7		32 7·8	31 5·0	2 0·3
4U	107 17·8	2 0·3		6 0·8	30 5·2	33 5·1		14 2·5	22 3·9
5R	78 13·4	2 0·2		10 1·4		41 6·3	11 1·6		14 3·9
5U	43 7·4		1 0·2	1 0·2	4 0·5	4 0·5	12 1·9	21 4·2	

* The figures in each row show to which positions the errors at a particular site of stimulation were referred. The totals columns give the total number of errors which occurred at each stimulus site. The actual errors are in roman type and the errors expressed as a percentage of the number of stimuli at each site are in italics. The hatched squares are those sites to which stimuli were correctly referred and at which therefore no error occurred.

Within these terms of reference two main types of error can occur : finger errors and side errors. There is no *a priori* reason why the occurrence of these two types of error should depend on the same factors and it was thought worth while to analyse them separately. For the purpose of this analysis finger errors are those errors in which the finger is incorrectly indicated regardless of the correctness or otherwise of the localization of the side of the finger stimulated. Similarly, side errors occur when the stimulus is referred to the wrong side of the finger whether the finger is correctly localized or not. The mutually inclusive case of "mislocalizaton" of both the finger and the side in the same response is not considered separately.

In Fig. 2 the distributions of finger errors and side errors are presented graphically in the form of histograms. It is at once apparent that patterns of distribution for the two types of error are markedly different. Both types of error are on the average

TABLE II

DISTRIBUTION OF ALL ERRORS* BEFORE AND AFTER OPERATION

Sites of Stimulation	Sites to which Stimuli are Erroneously Referred								χ^2	Degrees of Freedom	P (Level of Significance)
	2R	2U	3R	3U	4R	4U	5R	5U			
2R		78	76	8	37	5	13	5	204·1	6	<0·001
2U	137		62	109	40	35	22	4	238·4	6	<0·001
3R	81	26		61	155	18	16	1	337·6	6	<0·001
3U	4	27	135		88	298	11	11	837·8	6	<0·001
4R	13	6	140	30		62	62	6	410·8	6	<0·001
4U	4	0	13	59	123		35	46	273·3	6	<0·001
5R	5	0	12	0	70	17		32	192·8	6	<0·001
5U	0	1	1	4	6	30	51		174·5	6	<0·001

* The errors made on each hand are summed and presented as in Table I.

more frequent following stimulation of the ulnar side of a finger but in the case of side errors this ulnar predominance is much more marked. Summing the pre- and post-operative results (raw data) for both hands, ulnar side errors are in fact 2·05 times as frequent as radial side errors, whereas in the case of finger errors the proportion is 1·1 to 1. Again the incidence of the two types of error is different for the individual fingers, side errors occurring most frequently on the radial side of the hand whereas finger errors predominate on the two central fingers. A χ^2 test (Table III) shows that the distributions over the eight sites of finger errors and of side errors are different to a significant degree. In addition to the difference in frequency between ulnar side errors and ulnar finger errors there is also a marked difference between the distributions over the four fingers of the two types of ulnar error. Fig. 2 shows clearly that in the case of ulnar side errors there is a gradient of increasing weakness from the ulnar to the radial side of the hand. Although ulnar finger errors are also more frequent on the radial side of the hand, there is not the same gradient of weakness and the errors are distributed more with respect to the inside (centre) and outside (periphery) of the hand. This tendency for finger errors following stimulation of the ulnar side of a finger (ulnar errors) to increase towards the centre of the hand also characterizes radial finger errors and indeed is characteristic of finger errors as a group.

The remaining errors, radial side errors, are not distributed in a way which is obviously similar to the distribution of ulnar side errors. Like the finger errors they tend to be more frequent on the central fingers and, with the exception of the left hand post-operatively, they are more frequent on the ulnar side of the hand.

The observation that the factors governing the occurrence of side errors and finger errors appear to be different and that the differences in the distributions reach a high level of significance justify the

TABLE III

DIFFERENCE IN DISTRIBUTION OF SIDE ERRORS AND FINGER ERRORS*

	Site of Errors								Totals
	2R	2U	3R	3U	4R	4U	5R	5U	
Side errors ..	96	257	106	237	104	175	49	58	1,082
Finger errors ..	154	262	295	440	257	157	104	42	1,711
Totals ..	250	519	401	677	361	332	153	100	2,793

* These figures are obtained by summing the pre-operative and post-operative results for both hands for the 22 subjects. The difference between the distributions gives a value for χ^2 of 482·3, which, with 7 degrees of freedom, is highly significant ($P<0·001$).

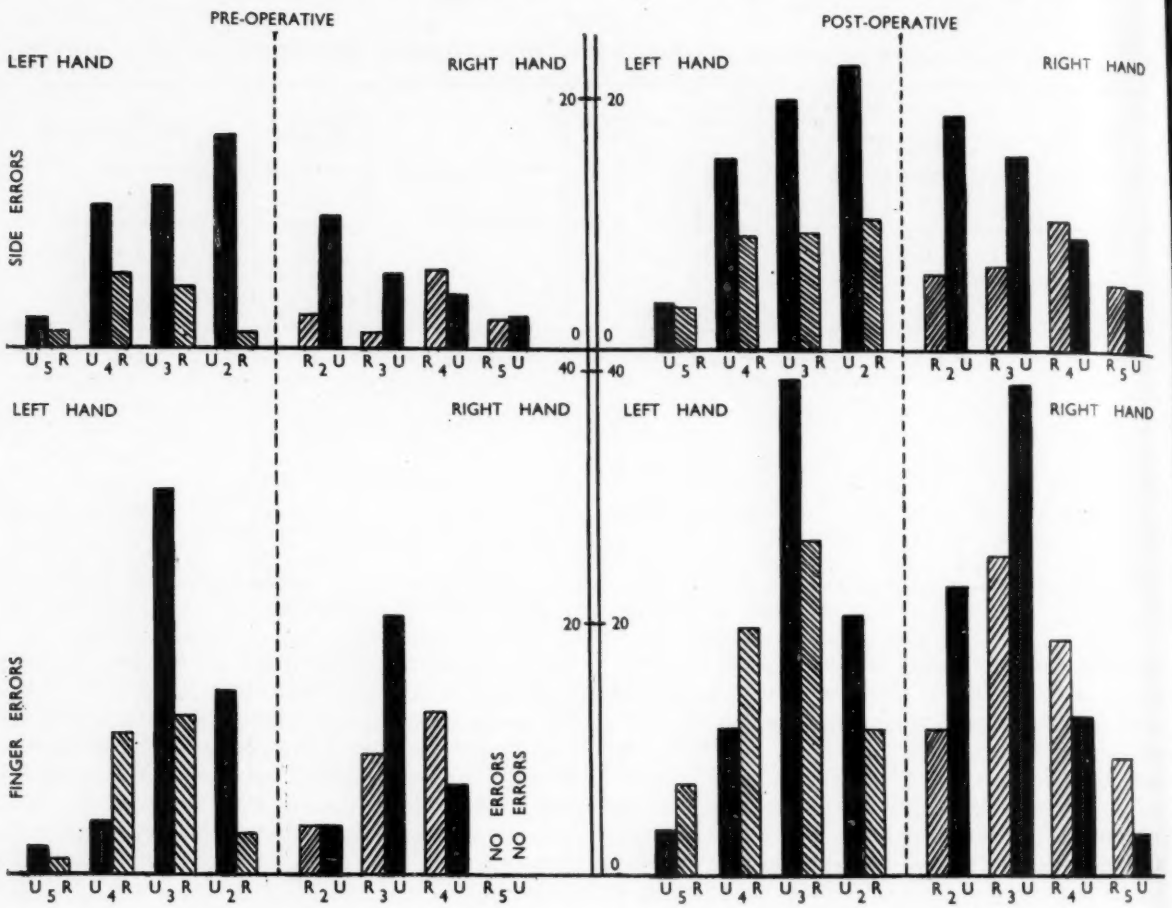

FIG. 2.—The incidence of finger errors and of side errors is given in separate histograms. The errors are shown for each side of each finger, both pre-operatively and post-operatively. Radial errors are hatched, those following ulnar stimulation are in black.

conclusion that the type of error which occurs most frequently (finger error or side error) is a function of the site of stimulation.

Direction of Errors.—The sites to which stimuli on the fingers are erroneously referred may also be considered with respect to the direction of mislocalization. This is not an independent variable since the opportunities for displacement in a given direction vary with the site of stimulation. Thus, in the case of side errors, stimuli on the ulnar side of a finger can only be referred to the radial side of a finger, and, in the case of finger errors, stimuli of digits 5 and 2 can only be referred in radial and ulnar directions respectively. Stimuli on digits 3 and 4 may be referred to fingers in either direction, but it is clear that there is a greater probability of a finger error following stimulation of digit 3 being referred in an ulnar direction than in a radial direction, and conversely for digit 4.

It has been pointed out above that in the case of side errors 67·2% of these errors follow stimulation of the ulnar side of a finger. This is equivalent to saying that 67·2% of side errors are referred in a radial direction since, in the case of side errors, direction of referral and the side of the finger on which the error occurs are interdependent.

In the case of finger errors 81·4% of such errors occurring on digit 3 and 36·0% of those occurring on digit 4 are referred in an ulnar direction. That is to say, in the case of both of the fingers in which there is some freedom in the direction in which finger errors may be referred there is a tendency for errors to be referred towards the middle two fingers of the hand. Analysis of Table I shows that there is an overall tendency for finger errors to be referred in an ulnar direction (68%) rather than in a radial direction. This cannot be accounted for by the excess of errors on digit 3.

A more striking result is the dependence which is

TABLE IV

CONTINGENCY BETWEEN DIRECTION OF FINGER ERRORS AND SIDE OF FINGER STIMULATED FOR FINGERS 3 AND 4*

Finger 3				Finger 4			
	Ulnar Stimulation	Radial Stimulation	Totals		Ulnar Stimulation	Radial Stimulation	Totals
Finger errors referred ulnarly	408	190	598	Finger errors referred ulnarly	81	70	151
Finger errors referred radially	31	107	138	Finger errors referred radially	72	189	261
Totals	439	297	736	Totals	153	259	412

* These figures are obtained by summing the pre-operative and post-operative results for the 22 subjects. Finger 3: $\chi^2 = 97{\cdot}6$, which, with 1 degree of freedom, is significant ($p<0{\cdot}001$). Finger 4: $\chi^2 = 27{\cdot}8$, which, with 1 degree of freedom, is significant ($p<0{\cdot}001$).

observed of the direction of referral of finger errors upon the side of the finger stimulated (Table IV). Errors following stimulation of the ulnar side of a finger are more frequently displaced in an ulnar direction while those following radial stimulation tend to be displaced radially. In the case of digit 3 a χ^2 test shows the contingency of direction of reference of finger errors upon the side of the finger stimulated to be significant at the 0·001 level of confidence ($\chi^2 = 97{\cdot}6$). In the case of digit 4 the test gives a result which is also significant at the 0·001 level ($\chi^2 = 27{\cdot}8$). The test cannot be applied in the case of digits 2 and 5 since in both these cases finger errors can be referred in only one direction, ulnarly and radially respectively.

It has been reported above that side errors following stimulation of the ulnar side of the fingers were much more frequent on the radial side of the hand and that radial side errors were slightly more frequent on the ulnar side of the hand. This fact may be expressed in terms of the direction of displacement of errors: the direction of displacement of side errors is contingent on the side of the hand stimulated, side errors on the fingers on the radial side of the hand tending to be displaced radially and conversely for side errors on the ulnar side of the hand.

These two effects, the contingency of the direction of finger errors on the side of the finger stimulated and the contingency of direction of side errors on the side of the hand stimulated, may be due to a "contamination" between two conceptual systems involved in the performance of the task. These two conceptual systems would relate to perception of the side of the hand stimulated and to perception of the side of the finger stimulated. Such evidence has implications for a theory of tactile local sign and will be discussed below.

Further Observations

Presentation of our observations in a summary manner tends to obscure the details of the errors made by our subjects. Thus in the immediate post-operative period one or two seriously confused patients made bizarre types of response. On one occasion one subject remorselessly indicated each finger in turn regardless of the site of stimulation. For a short time post-operatively several patients made errors of a perseverative type, repeatedly indicating the same position although successive stimuli were at different sites. It was also apparent that subjects would occasionally confuse the two sides of the hand. Following stimulation of the second finger, for example, a subject would hesitate between the two sides of the hand but show no interest in the middle two fingers. It was also clear that mislocalization was not always related to a feeling of uncertainty. Thus for several sessions a subject would place the site of a particular stimulus incorrectly but with complete confidence and with no tendency to make a correct response.

Individual differences in error pattern were quite marked post-operatively, but as only six subjects made 10 or more errors each pre-operatively, it was not usually possible to establish that these individual differences existed before operation. In order to elucidate this point an alternative method of investigating tactile localization has been devised and the results will be reported in a subsequent communication.

In using the method of Henri we have assumed that the localization of stimuli to a three dimensional model is a task which is essentially similar to the localization of stimuli to the actual site of stimulation on the hand of the subject. Both of these tasks involve matching in the sense in which Head employed the term, and this, he claimed, did not involve verbalization. It seems likely that the difference between the two tasks is in the degree of difficulty rather than in the nature of the functions involved. Observation of a few of our patients showed that, even when the "groping error" is excluded, localization to the subject's own hand was a more accurate performance but errors of a

"conceptual" type were still present. Thus different distributions of errors at the different sites were observed in the case of finger errors and side errors, as were some of the contaminatory effects discussed above. Differences between the results obtained using the two methods did occur but these differences were not such as might alter the theoretical position maintained in this paper with respect to errors made in localizing to a model. It is hoped to report in a later paper differences due to variations in technique and their implications for a theory of tactile localization.

Effect of the Operation

In the main analysis little distinction has been made between the results obtained pre-operatively and those obtained after leucotomy. This has been justified by the consistency of the results obtained and the close similarity of pre-operative and post-operative error patterns. Nevertheless, differences do exist. It has been reported above that the total percentage of errors increases following the operation. In addition to this there are some changes in the distribution of errors on the hands. In the case of the left hand (Table V) a χ^2 test shows

TABLE V
DISTRIBUTION OF ERRORS ON THE LEFT HAND FOLLOWING LEUCOTOMY*

	Sites of Incidence of Errors							
	2R	2U	3R	3U	4R	4U	5R	5U
Pre-operative ..	7	31	22	53	17	16	4	4
Post-operative ..	120	186	169	260	128	143	50	43

* The figures shown in this table are the total errors made on the left hand by the 22 subjects. $\chi^2 = 15{\cdot}3$, which, with 7 degrees of freedom, is significant ($p < 0{\cdot}05$).

that the post-operative distribution of errors on the eight sites is different from the distribution observed pre-operatively at the 0·05 level of significance ($\chi^2 = 15{\cdot}3$). The same test applied in the case of the right hand gives an insignificant value for χ^2. That is to say, there is no difference between the pre- and post-operative distributions of total errors on the right hand which is not easily attributable to chance. Inspection of the original data suggests that the change in the left hand is accounted for largely by a post-operative decrease in the number of side errors following stimulation of the ulnar side of a finger in the left hand. It is noteworthy that the effect of this decrease is to make the distribution or errors on the two hands more similar post-operatively than was the case pre-operatively. Thus, when errors are expressed as the percentage of the number of stimuli, post-operatively the rank order of the different sites with respect to error frequency is identical in the two hands, whereas pre-operatively there are six discrepancies in rank order, one site differing in the two hands to the extent of three positions of rank order.

Unfortunately, when the data under consideration are the results summed for all subjects, unequivocal changes in the distribution of errors following operation cannot be proved. This is precluded by the possibility that individual subjects with different error patterns may be unequally impaired by the operation and so produce an artifactual change in the mean pattern of errors.

It is not proposed to discuss individual differences in detail here, as this question will be reported upon elsewhere. However, it may be said that our results suggest that, in at least some of our subjects, the operation produces a change in the distribution of errors. Thus, in two of our subjects it can be shown that pre-operatively the ratio of errors following ulnar stimulation of a finger to those following radial stimulation is greater in the left hand than in the right, and that this difference is significantly reduced following the operation. It is possible that, in these cases, pre-operatively the side of the finger stimulated, left or right, is being used as a standard of judgment and that post-operatively this dimension of orientation is less prominent. This would be consistent with the results of the data summed for all subjects in so far as they suggest that frames of reference other than the anatomy of the area stimulated may play a role in tactile localization. Orientation in terms of left and right may be such an "external" frame of reference.

Discussion

The problem of tactile localization has two main aspects which present different but interrelated problems. First, the question may be asked, How much and what type of information is conveyed to the central nervous system? Secondly we may ask, What are the mechanisms of analysis which enable an appropriate localizing response to be made? This is the problem which led Head to postulate a dynamic scheme of orientation which was continuously modified by incoming stimuli. Recognition of the problem of this kind of organization is implicit in the term "body image", a term which, although it has teleological reference, is generally used to connote an unexplored region of neurological function.

It has been shown here that gross errors of tactile localization occur in the absence of neurological lesions, and that following prefrontal leucotomy,

although the number of errors is increased, the post-operative breakdown of the skill is very similar to the pre-operative error pattern.

It seems likely therefore that our results represent the patterns of weakness inherent in a highly complex skill constituting normal tactile localization rather than an agnosia resulting from a complete deficit of a perceptual function, such as occurs in parietal lesions. Our results suggest that the perceptual responses involved in " local sign " are made in accordance with certain definable dimensions of orientation. Thus, if the distribution of errors is considered as a whole it appears haphazard, but if the errors are considered according to whether the type of error involved is a finger error or a side error the distributions fall into patterns which have obvious conceptual correlates. Thus finger errors increase from the outside of the hand to the inside while side errors occur principally on the ulnar side of the digits, and these ulnar side errors are much more frequent on the radial side of the hand.

Of particular interest is the contingency of the type and direction of error on the site of stimulation. Thus the different distribution on the hands of finger errors and side errors suggests that the subject may decide separately which finger and which side of a finger is stimulated. This observation would be difficult to explain as a failure of a directly representational system, and even less consistent with such a theory are the observations concerning the effect of site of stimulus on the direction in which stimuli are erroneously referred. Thus finger errors are displaced mainly in the direction of the side of the finger stimulated, and radial and ulnar displacement of side errors most frequently follow stimulation of fingers on the radial and ulnar sides of the hand respectively. Such results are more consistent with the hypothesis that localizing responses to tactile stimuli are made on the basis of a group of concepts in terms of which such tactile stimuli are organized. According to this theory, when errors occur their location is likely to be determined not only by the adequacy of sensory information but also by the efficiency of interpretation. Moreover, the direction of displacement of errors could be partly determined by a confusion between two frames of reference. Thus, a stimulus on a finger on the ulnar side of the hand would have a quality of " ulnarness " with respect to the side of the hand stimulated, and this quality might wrongly be attributed to the side of the finger stimulated. That is to say, the frame of reference by which the side of the finger stimulated is recognized is contaminated by the frame of reference by which the side of the hand stimulated is recognized. In the same way, a stimulus on the ulnar side of a digit may erroneously be referred towards the ulnar side of the hand, as is also found to occur in our results. In this case, the awareness of " ulnar " stimulation derived from the side of the finger stimulated is attributed to the side of the hand. This kind of effect could be described as the confusion of two dimensions of orientation. " Conceptual contamination " of this type has been observed experimentally, particularly in sorting tests of conceptual thinking or " categorical abstraction ". For example, in Weigl's (McFie and Piercy, 1951) test an organically impaired patient may attempt to sort objects by their shape but the performance may be contaminated by a tendency to sort according to colour at the same time.

In some of our subjects the ratio of errors following ulnar stimulation of a finger to those following radial stimulation is pre-operatively greater in the left hand than in the right. This difference suggests the possibility that the side of the finger, left or right, is also used as a standard of judgment. However, the data is insufficient or the effect too small to stand up to tests of statistical significance in more than two of our subjects. It is noteworthy, however, that the effect is reduced post-operatively. Such an impairment of a dimension of orientation would be consistent with the restriction of perceptual grasp commonly observed in cases of organic brain damage.

Tactile localization on the fingers may thus be regarded as a special instance of the general problem of orientation, and it may be that the orientation of an individual in relation both to his own body and to his external environment depends upon the integrity of a comparatively small number of simply structured dimensions of reference which determine the interpretation placed on incoming stimuli.

Summary

Tactile localization on the fingers has been studied pre- and post-operatively in patients undergoing prefrontal leucotomy. Although the frequency of gross errors was increased post-operatively similar errors were made before operation, and it was concluded that the operation impaired an imperfect skill in an essentially non-specific manner. The incidence of errors and the direction in which stimuli were erroneously referred were found to depend on the site stimulated. Thus the distribution of errors resulting from the subject's indicating the wrong finger (finger errors) was different from the distribution of errors resulting from the subject's indicating the wrong side of a finger (side errors).

This suggests that the subject decides separately which finger and which side of a finger is stimulated. Further, the direction of referral of finger errors is positively related to the side of the finger stimulated, and the direction of referral of side errors is similarly related to the side of the hand (ulnar or radial) stimulated. This result has been interpreted as a confusion on the part of the subject between two standards of reference or " concepts ", the side of the finger stimulated and the side of the hand stimulated.

These observations are consistent with the hypothesis that normal tactile localization in man occurs through the intermediary of certain dimensions of orientation or concepts. The relevance of these observations to theories of postural orientation and orientation in external space is indicated.

The subjects for this investigation were patients under the care of the physicians and surgeons to the National Hospital, Queen Square, London, and we wish to express our thanks for their permission to study their patients. To Dr. E. Arnold Carmichael, Dr. G. D. Dawson, Dr. Eliot Slater, and Professor O. L. Zangwill we are indebted for criticism and advice.

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BOOK REVIEWS

Lehrbuch der Analytischen Psychotherapie. By H. Schultz-Hencke. (Pp. 340. DM. 27.–.) Stuttgart: Georg Thieme. 1951.

The only recognized way of studying psychoanalysis as a therapeutic technique is by the student submitting himself to a personal teaching analysis. The need of a textbook of analytic psychotherapy has, therefore, not been felt to be a pressing one in England and the U.S.A. The publication of a textbook could, indeed, be regarded by some analysts as a heretical venture, as it might encourage some psychotherapists to embark on treatment of this kind, after study of the book, without any personal indoctrination. The author's heresies do not end at this point. He has also attempted to provide in his textbook a synoptic version, in which the several contributions of Freud, Adler, and Jung are given their own place, and are worked into a harmonious whole. Orthodox Freudians will not regard this with approval. Other ways in which Dr. Schultz-Hencke's exposition differs from generally accepted standards are in insisting on a monistic solution of the body-soul problem, in giving some appreciation of genetical and constitutional factors in the causation of neurosis, and in describing the physical and physiological pathways along which symptoms may be shown. The general psychiatrist will welcome the breadth and liberality of mind which has informed this undertaking, and the interesting attempt which has been made to introduce a scientific eclecticism into a branch of psychiatry which has tended to become doctrinaire.

Morbus Alzheimer and Morbus Pick, a Genetic, Clinical and Patho-anatomical Study. By T. Sjogren, H. Sjogren, and A. G. H. Lindgren. (Acta Psychiatrica and Neurologica Supplement 82.) (Pp. 152. Sw. Cr. 25). Copenhagen: Munsgaard. 1952.

This book maintains the high standards of general production that one associates with *Acta Scandinavica* supplements. It is an English translation, and though occasionally lacking polish, the meaning is always clear. Based on the records of 80 cases, 18 each of histologically verified Alzheimer and Pick disease, and 43 with a clinical diagnosis of Alzheimer-Pick syndrome but without any histological proof, the material is reviewed in its genetical, clinical, and pathological aspects.

In the first section the influence of a hereditary factor is clearly shown, and the suggestion made that multifactorial inheritance is more likely in Alzheimer's disease, while a dominant major gene with modifying subordinate genes seems more probable in Pick's disease.

On the clinical side the difficulties of differential diagnosis are made clear. Both show changes of conation early, and later various amnesic and dysphasic symptoms. The old psychiatric mnemonic "placid Pick, anxious Alzheimer" is not borne out in these records, though Parkinson-like difficulties in gait, and increased muscle tone seem, in this series, sufficiently more frequent in the Alzheimer cases to be of diagnostic value.

In the sphere of pathology the clear-cut differences noted by earlier writers is confirmed. Alzheimer cases show diffuse, widespread outer layer cortical changes, especially of frontal and temporal lobes, Pick's show circumscribed, though often multiple, areas of change involving the whole depth of cortex. The characteristic differential cytological changes in the two are also confirmed. One addition to previous work is the high proportion of Alzheimer cases showing changes in the basal ganglia and below.

This careful and detailed study is a useful contribution both to knowledge of the Alzheimer-Pick syndromes, and to the more general problem of the senile and presenile psychoses.

Personality and the Frontal Lobes. By Asenath Petrie. (Pp. 188. 21*s*.) London: Routledge & Kegan Paul. 1952.

Mrs. Petrie studied psychology in Dr. Eysenck's department at Mill Hill Emergency Hospital, London, and at the Maudsley Hospital, London, and when she went to work in the Department of Psychiatry at St. George's Hospital, London, she employed the techniques she had learned in a non-surgical environment to the results obtained by neurosurgery.

Twenty-seven patients were subjected to psychological tests both before and after leucotomy. The patients suffered from affective and neurotic conditions, e.g. obsessional neurosis, and only one of them is said to be worse after operation. A very large battery of tests was given, covering, apart from intellectual functions, two of the "dimensions of personality" formulated by Eysenck, "neuroticism" and "introversion". The statistical analysis is carried out by subtracting the patient's pre-operative from his post-operative score, and showing that the difference varies significantly from zero. The disadvantage of this presentation is that one is unable to know whether the change is a small or a large one absolutely. It is of little value to be told that three months after operation the patients showed a mean diminution of 2·35 points in suggestibility, unless one knows what are the mean scores and standard deviations for normals and for neurotics on this test.

On the average, patients tended to show lower scores after operation both in respect of "neuroticism" and "introversion". They also had lower scores to tests of verbal and full-scale I.Q. Though interesting, these results will not be of much positive value to the clinician. More would have been obtained, even from this stati-

stically small material, if the collaboration of a clinician had been secured, and individual test results had been correlated with degrees of clinical improvement.

It is to be regretted that the author did not publish the results of her research as a series of communications to the psychiatric press. Production in book form is not well suited to this type of report, as much unnecessary matter, such as an account of the standard leucotomy operation, is included.

Meconnaissances et Hallucinations Corporelles. By H. Hécaen and J. de Ajuriaguerra. (Pp. 370; 35 figures. Fr. Fr. 3,200.–) Paris: Masson et Cie. 1952.

Once again these authors present an admirably comprehensive collection of case histories illustrating one of the spheres of interaction between mind and brain. The cases, some of which are quoted from the literature while others are reported for the first time, cover disturbances of the body image in whatever form it may be found, from phantom limbs to psychotic depersonalization states. The psychological, physiological, and anatomical disturbances accompanying the symptoms are analysed, not in order to argue any one particular hypothesis but in a spirit of critical appreciation. After a final brief survey of the patterns of development and disintegration of the body image, the authors conclude that no single factor is responsible for its maintenance, but that it is the result of an organization of many activities and influences, including social and biological forces as well as sensory-motor impulses.

It is not so much for any originality of thought or approach that this volume will be welcomed, but because the amount of information it contains and its clarity of exposition makes it an invaluable textbook for those who wish to investigate the subject further.

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(Review in a later issue is not precluded by notice here of books recently received.)

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History of American Psychology. By A. A. Roback. (Pp. 426; illustrated. $6.00.) New York: Library Publishers, Inc. 1952.

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